RESIDENT/FELLOW RESEARCH
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A Comparison of Antibiotic Use Appropriateness Between High and Low Prescribers in the Baltimore Veterans Affairs Medical Center Outpatient Setting
Identification of Primary Care Patients at Risk of Non-Alcoholic Steatohepatitis (NASH) with Advanced Fibrosis Using the NAFLD Fibrosis Score and Abdominal Ultrasound Imaging

Authors: Namphuong T. Tran, MD; Alissa Detz, MD; Edward W. Holt, MD

Introduction: Non-alcoholic fatty liver disease (NAFLD) affects 80 million Americans and is associated with the metabolic syndrome. Patients with NAFLD and non-alcoholic steatohepatitis (NASH) can be clinically asymptomatic as they progress to cirrhosis; early identification is essential for preventing disease progression. We evaluate the identification of patients at highest risk for NASH based on the validated NAFLD Fibrosis Score (NFS) in a primary care setting, and further steps taken toward workup, including correlation with radiographic evidence of fibrosis.

Methods: We retrospectively reviewed the medical records of adults 50 years and older, diagnosed with diabetes, seen between 2013 and 2016. Patients were included with alanine aminotransferase (ALT) levels above the upper limit of normal (40 U/L), and with other sufficient clinical data to calculate the NFS. Patients were excluded if they had ALT levels of greater than 400 IU/L, chronic hepatitis B or C infection, as well as alcohol consumption of greater than 14 standard drinks per week (women) or greater than 21 drinks per week (men). Patients were designated at high risk for NASH with advanced fibrosis (F3-4) if the NFS was >0.655, indeterminate risk for NFS 1.455 - 0.655, and low risk for NFS < 1.455. Ultrasound features considered concerning for advanced fibrosis were portal vein size > 10mm, splenomegaly and the presence of hepatic parenchymal heterogeneity.

Results: Our review identified 847 patients at higher risk for NASH with fibrosis. After exclusions, our cohort consisted of 110 patients: 56 males and 54 females. The mean age of the study cohort was 69 +/- 1.7 years, the average BMI was 27.31 +/- 3.5 kg/m2 and the average ALT was 70.1 +/- 28.6 U/L. 29 patients (26.3%) were identified as low risk of advanced fibrosis, 65 (59.0%) indeterminate risk and 16 (14.5%) high risk. Abdominal ultrasounds were obtained for 4 (13.7%) of the low risk patients, 8 (27.5%) of the indeterminate and 17 (58.6%) of the high risk patients. Of those ultrasounds obtained, 0, 7 (24.1%) and 22 (75.9%) in the low, indeterminate and high risk groups respectively demonstrated features concerning for advanced fibrosis. Only 9 patients (4 low, 2 indeterminate, 3 high risk) were referred to a hepatologist.

Conclusion: Patients at risk for NASH appear to be considerably under-recognized in the primary care setting. The NFS is a simple, validated clinical equation that can help identify patients at risk. Higher risk patients who undergo abdominal ultrasound may have correlating radiographic features of otherwise asymptomatic advanced fibrosis, further demonstrating the importance of diligent recognition of risk. Efforts to identify these patients at highest risk in primary care could lead to earlier identification and treatment of patients and ultimately reduce the growing complications of advanced disease.
Giant Cell Arteritis: Not Just a Disease in Caucasians

Authors: Cassie Xu, MD; Thomas Bush, MD

Introduction: Giant cell arteritis (GCA) is seen mainly in Caucasians of Northern European descent, and its presentation, treatment, and prognosis have been well described in this population. But existing epidemiologic studies are limited because they have been performed on predominantly Caucasian populations. This research study fills a gap in the literature by examining GCA in a healthcare setting that includes a large Hispanic population. It hypothesizes that the incidence and presentation of this disease in the Hispanic population is similar as in non-Hispanic populations.

Methods: We performed a retrospective-chart review on patients seen in Santa Clara Valley Medical Center, a county hospital in San Jose, California. Approximately 35 percent of patients in our system identify themselves as Hispanic. We conducted a search of our electronic medical record (Epic) from 2008 to 2017 and identified 17 patients with a diagnosis of GCA—10 were biopsy proven and 7 were diagnosed on clinical grounds by rheumatologists. This study describes the disease presentation, diagnosis, and treatment for these patients.

Results: Eight of the 17 patients with GCA were Hispanic (47%). The average age of Hispanic GCA patients was 80, with an average diagnosis age of 71. In comparison, the average age of non-Hispanic GCA patients was 78, with an average diagnosis age of 69. The two groups presented similar symptoms, the most common being headache, jaw pain/discomfort, and vision loss. Additionally, all the patients had imaging done, some as part of their GCA diagnosis/surveillance and others as part of unrelated presentations and hospital visits. Two of eight Hispanic patients (25%) and two of nine non-Hispanic patients (22%) had large-vessel involvement per imaging. These results are similar to some past studies, which have shown a 10-15% involvement of aortic arch and 26% for subclavian and axillary arterial involvement.

Conclusion: In conclusion, this study indicates that the prevalence of GCA in Hispanic populations is similar to that of non-Hispanic populations. This suggests that the historical view of GCA as mainly affecting Caucasian populations of Northern European descent might need to be broadened to encompass other populations.
Computed Tomography Fractional Flow Reserve: An appropriate low-risk screening tool for coronary disease

Authors: Alexander Robinson DO; Nicholas Isom MD; Christopher Buckley DO; Thomas Rosamond MD

Introduction: Left heart catheterization for direct visualization of coronary vessels has been common practice for many years. The decision to perform percutaneous coronary intervention (PCI) is often based upon the observed percent stenosis in each vessel, and vessels with 70% or greater stenosis often have intervention performed. Over the last decade, fractional flow reserve (FFR) has gained traction in determining if a lesion is hemodynamically significant [1]. FFR uses direct measurement of pressure and flow to determine if the stenosis is truly causing significant ischemia—thus giving a more approachable and objective measurement to assist with making the decision to intervene[2]. More recently Computed Tomography fractional flow reserve (CT FFR) imaging has allowed physicians to obtain an FFR value without requiring an invasive left heart catheterization. As this is a relatively new technique, there is limited data comparing CT FFR with direct visualization of left heart catheterization.

Methods: 71 patients received CT FFR during their care at Kansas University. Of those 71 patients, 19 patients had a diagnostic left heart catheterization as part of an ischemic workup. Seven of those patients had to be excluded due to misalignment, motion artifact, or previous stents obscuring the results. The 12 remaining patients had their CT FFRs compared with their catheterization results. An FFR result of less than 0.8 was considered to be hemodynamically significant, while a stenotic lesion of 70% or more was also determined to be significant.

Results: Using the guidelines noted previously for determining significant lesions, five of the patients were found to have FFR values that were less than 0.8. Three of those patients were then found to have significant stenosis on catheterization. The remaining seven patients receiving CT scans all had non-significant FFR values. All seven of those patients had negative left heart catheterizations as well. For purposes of screening or diagnostics, CT FFR was found to have a sensitivity of 100% and specificity of 77.8% when compared to gold standard left heart catheterization.

Conclusion: Using the guidelines noted previously for determining significant lesions, five of the patients were found to have FFR values that were less than 0.8. Three of those patients were then found to have significant stenosis on catheterization. The remaining seven patients receiving CT scans all had non-significant FFR values. All seven of those patients had negative left heart catheterizations as well. For purposes of screening or diagnostics, CT FFR was found to have a sensitivity of 100% and specificity of 77.8% when compared to gold standard left heart catheterization.
Introduction: Patients evaluated after sexual assault may require non-occupational post-exposure prophylaxis (nPEP) to prevent infection with human immunodeficiency virus (HIV), depending on the assessed risk of HIV transmission in each case. Access to nPEP medications, patient counseling, and follow-up care should be offered in a systematic, comprehensive, and compassionate setting. Unfortunately, multiple barriers may impede this process. The University of New Mexico (UNM) IN-STEP (Integrating nPEP after Sexual Trauma in Emergency Practice) project is a trainee-driven, multidisciplinary, interdepartmental quality improvement (QI) effort to improve HIV prevention in patients evaluated after sexual assault, while emphasizing the centrality of the patients’ experience in the resulting process of care.

Methods: The IN-STEP team identified and addressed several key areas for clinical QI and infrastructure development. An emergency department (ED) nPEP prescribing algorithm was developed. Funding was secured from the UNM Committee of Interns and Residents QI grant program to cover the full treatment cost of nPEP medications for patients evaluated after sexual assault. Patient and provider education materials were developed in collaboration with IN-STEP team members and the New Mexico AIDS Education and Training Center. An ED provider survey was conducted to inform project planning and provider education. A parallel-cycle Plan-Do-Study-Act (PDSA) analysis was used to track the complex, concurrent QI efforts undertaken in each area of the project; and an IN-STEP dashboard was developed to facilitate project communication.

Results: Four key areas for improvement were identified. These included: (1) access to HIV testing in the ED; (2) provision of nPEP medications, using a patient-centered approach; (3) continuity of care between the ED and 10 follow-up sites within the community; and (4) education and training of ED and community site providers. These key areas corresponded well with the barriers to nPEP delivery identified by surveyed ED providers (n=42). PDSA cycles were prepared for each key area, and a composite cycle was shared with other stakeholders at the institution. The IN-STEP dashboard was a useful tool for project communication. IN-STEP was instrumental in implementing ED point-of-care HIV testing, an ED clinical workflow with nPEP decision support, nPEP medications available at no cost for patients evaluated after sexual assault, numerous patient educational materials, and access to follow-up care coordinated through a 24/7 phone line.

Conclusion: The infrastructure developed for IN-STEP resulted in significant systems improvements in HIV screening, prevention, and continuity of care at our institution, influencing the care of patients affected by sexual assault as well as those evaluated for other indications. These results support the implementation of complex QI efforts using parallel-cycle PDSA analysis and highlight the importance of implementing such efforts with a multidisciplinary team. Lessons learned from this project may be useful for other large-scale, multidisciplinary efforts.

References
Neoadjuvant Chemotherapy in Triple Negative Breast Cancer and Its Impact on Tumor Progression and Overall Survival: A Tertiary Care Center Experience.

Authors: Johnny Chahine, MD; Bicky Thapa, MD; Oscar Perez Gomez, MD; Hamed Daw, MD; Abdo Haddad, MD., Cleveland Clinic, Fairview Hospital, Cleveland, Ohio, USA.

Introduction: The incidence of triple negative breast cancer (TNBC) is about 13% as per recent national cancer database analysis and it is aggressive cancer with limited treatment options. Systemic chemotherapy, surgery and radiation remains a mainstay of therapy. Pathological complete response (pCR) has been observed as an important prognostic factor with better survival benefits in patients with TNBC.

Methods: After IRB approval, we retrospectively reviewed TNBC cases from 2008-2014 in Cleveland Clinic Database. Patients who received Neoadjuvant chemotherapy were assessed for local and distant progression using univariable and multivariable competing risk analysis.

Results: A total of 156 patients with TNBC treated with neoadjuvant chemotherapy was identified. Mean age at the diagnosis was 53.78, 119 (77.3%) were white, 33 (21.4%) black. Most of the patients had ECOG (Eastern Cooperative Oncology Group) score of 0 (65.8%) and 1 (30.3%), BRCA was positive in about 18%, 124 (90.5%) had histologic grade 3 and 13 (9.5%) with histologic grade 2. Ductal histology was found in 126 (81.8%) 3 (1.9%) lobular, and 3 (1.9%) had mixed histology; surgical margin was positive in 14 (12.8%), vascular invasion was present in 43 (41.3%), and 48 (44.9%) had a lymphatic invasion. Sentinel lymph nodes (SLN) were positive in 60 (48.8%) and 58 (98.3%) underwent axillary node dissection. Number of patients in stage I, II, III, and IV were 9 (6%), 71 (47.3%), 58 (38.7%) and 12 (8%) respectively. About 151 (98.1%) underwent surgery, out of which 122 (80.8%) had mastectomy and rest of the patients had lumpectomy; 120 (78.9%) received radiation therapy; with regards to chemotherapy 124 (80%) received AC-T (doxorubicin, cyclophosphamide and paclitaxel), 14 (9%) received TAC (Docetaxel, Doxorubicin, Cyclophosphamide), and 5 (3.2%) got TC (Docetaxel, Cyclophosphamide). Sixty-four patients had a relapse in the whole cohort, 49 achieved pCR (out of which 15 had the relapse).

Univariable analysis with competing risk analysis for both local and distant progression was significant for SLN positivity [Hazard ratio (HR) of 2.52 (1.44, 4.40), p = 0.001] and overall staging, stage I [HR 1.80 (1.25, 2.60) (p = 0.037)]. However multivariable analysis with competing risk analysis for both local and distant progression was only significant for overall staging of the TIBC [HR 2.08 (1.05, 4.13) (p = 0.037)]. Five-year Overall survival (OS) rate for stage I, II, III and IV TNBC were 86%, 80%, 40%, and 19% respectively.

Conclusion: Our analysis shows overall staging and SLN positivity significantly correlated with both local and distant progression, however SLN positivity failed to reveal any significance on multivariable competing risk analysis. So more effective therapy for TNBC patients still remains the unmet need for better survival and quality care.
IMPROVING RETINOPATHY SCREENING WITHIN THE COMMUNITY PRACTICE CENTER THROUGH THE USE OF TELERETINAL SERVICES

Authors: Lorena Rasquin, Karla Curet, Anna Lo, Horacio Hares, Mathew Behme

Introduction: Diabetic retinopathy is a leading cause of blindness in adults. In the U.S. less than 60% of diabetic patients undergo proper screening. The Community Practice Center (CPC) at Albert Einstein Medical Center serves one of the most complex neighborhoods in Philadelphia with a large racial/ethnic minority population with multiple co-morbidities and social determinants of health. The current standard of care for diabetic retinopathy screening falls below the 30th percentile for HEDIS measures according to quality gap report by Health Partners Plans (HPP). Our aim is to increase the rate of retinopathy screening within one year of implementation by incorporating the RetinaVue imager to the Diabetes clinic.

Methods: The CPC started a comprehensive patient-centered model of diabetes management visits where clinicians perform diabetic foot exam, nephropathy screening, blood pressure measurement, point of care hemoglobin A1c, and education. As a quality improvement project, we incorporated retinopathy screening with Welch Allyn RetinaVue 100 imager instead of ophthalmology referral. This device captures nonmydriatic retinal image which is uploaded and evaluated by an ophthalmologist. PDSA cycles where performed every 5 weeks to improve our image capture rates and streamline workflow. We compared data from patients in the diabetes clinic before and after incorporating the device.

Results: Before implementation of the device 69 patients were seen in the diabetes clinic, 47 were pending retinopathy screening and 14 completed their screening at their next appointment within 6 months.

Preliminary data after 9 months of implementation with the device 85 patients were seen in the DM clinic, 69 were pending retinopathy screening, all had imaging attempted, and 38 were successful. Those with unsuccessful images received ophthalmology referral.

In the pre-intervention group 40.9% of patients completed screening, as compared to post-intervention group where 59.1% completed retinopathy screening in office without the need of referral, increasing the screening rate by 18.2 percentage points (p=0.262, RR of 1.17).

Conclusion: This project represents an innovative patient centered approach to diabetes care in a training program where neuropathy, retinopathy and nephropathy screening can be performed in real time. This example of a population health management initiative illustrates the concepts of quality improvement to all trainees in the program. Our patient population experiences significant barriers to care leaving screening measures often abandoned for other urgent needs. Teleretinal service reduces barriers to care, allowing to provide better care. Result are not statistically significant and limiting factors include small patient sample, and learning curve for image capture. Continuous evaluation of capture rates, and staff training are essential for program success. Moving forward expanding the services to all patients in the clinic in addition to the diabetic clinic might increase our impact in screening rates.

Disclosure: We do not have any financial association with Welch Allyn.
Unnecessary Telemetry Utilization – A Multidisciplinary Approach to Reducing Healthcare Spending

Authors: Keel T, Odeti S, Vomer R

Introduction: The cost of healthcare in America is non-sustainable. Telemetry monitoring in the non-intensive care setting continues to be cited as one of the most overutilized diagnostic tests in hospital-based medicine, contributing directly to increased cost of hospitalization. Additionally, inappropriate telemetry monitoring often leads to a penumbra of unnecessary diagnostic and therapeutic interventions, further consuming house and staff resources.

Objective: To evaluate the effectiveness of systems-based and educational interventions in reducing unnecessary telemetry utilization.

Design: Our study was a single-center prospective study examining inpatient telemetry utilization for 1 year (July 2017-June 2018). Our approach was multifaceted, incorporating both education and timely clinical reminders. In addition to making posters outlining guidelines for appropriate usage readily available at all nursing stations, point-of-care guideline-based clinical decision-making support systems (CDSS) were incorporated into the EMR telemetry order set. Additionally, a standard of work was established wherein telemetry technicians were prompted at 24-hour intervals to remind clinicians to reexamine the continued need for telemetry.

Setting: A 130-bed academic hospital and community-based regional referral center in Southwestern Virginia.

Participants: All patients for whom telemetry monitoring was ordered during the study period were included in the study. Patients in the Intensive and Progressive Care Units were excluded on the grounds that they require advanced monitoring by hospital policy.

Main Outcomes: The primary outcome measured was the rate of telemetry utilization, as expressed by percentage of patients on telemetry. Secondary endpoints included both absolute and relative amounts of telemetry utilization (expressed in total hours of telemetry used, and days of telemetry per total number of patient days, respectively).

Results: By the end of the study, average telemetry utilization was reduced from 37.89% to 16.39% of hospitalized patients. Total amount of monitoring decreased from 501 hours per month to 195, translating to $16,740/month reduction in direct costs-of-care.

Conclusion: Our study has shown that simple, systems-based and educational interventions are effective at reducing unnecessary telemetry utilization. It was pragmatic in that it employed intervention-specific guideline-directed education and CDSS, coupled with easily integrative procedural interventions, as such we feel it is highly generalizable to various metrics at hospital systems at large. Our study was limited in that it was a conducted at a single-center community-based hospital. It was also limited in that, due to its pragmatic design, precise tracking of tertiary endpoint improvements (i.e., reducing unnecessary further testing and procedures) was impossible, thus reducing the accuracy of any extrapolated cost-savings. Additionally, our CDSS implementation was specific to our EMR, we are uncertain as to the ease with which such interventions could be applied in other systems. Nevertheless, we feel that our study effectively demonstrates that simple, education and systems-based interventions are effective at reducing unnecessary telemetry utilization.
EARLY-ONSET COLORECTAL CANCER HAS UNIQUE CLINICAL CHARACTERISTICS AND MOLECULAR FEATURES

Authors: Rawan Dayah, MD; Mohammad Bilal, MD; Nattaporn Tun, Tewfeek Abu-Shami, Shailendra Singh, Adam Booth, MD; Praveen Guturu, MD

Introduction: The incidence of colorectal cancer (CRC) in young patients has been increasing. Recently, the American Cancer Society (ACS) issued a qualified recommendation to start CRC screening at age 45. Despite this, early recognition of CRC in young patients continues to be a challenge. We aimed to evaluate the disease characteristics of patients with early-onset CRC (<50 years of age) and compare them with those of older patients (>50 years of age).

Methods: A retrospective cross-sectional study was performed using our electronic pathology database. All patients diagnosed with CRC between January 2012 and September 2018 were included. Data regarding patient demographics, comorbid conditions and patient presentation was noted. We also noted information regarding the location, staging and molecular features of CRC.

Results: A total of 627 patients with CRC were identified. Out of these, 117 (18.6%) were younger than 50 years of age (early-onset CRC). The mean age was 64.6 years and 41.1 years in patients > 50 years and < 50 years of age, respectively. Increased number of Caucasians had CRC after 50 years of age (59.6% vs 49.6%, p-value: 0.04) as compared to those <50 years of age, while significantly increased number of Hispanics had early-onset CRC (29.1% vs 17.2%, p-value: 0.02). The most common presenting complaint in both groups was rectal bleeding [Table 1]. Significantly increased number of patients with early-onset CRC presented with lower abdominal pain (37.6% vs 22.4%, p-value =0.001). Rectal cancer was more common in patients with early-onset CRC (32.5% vs 23%, p-value: 0.03), while more patients > 50 years of age had CRC in the ascending colon (20.7% vs 4.3%, p-value: 0.001). There were no significant differences in presence of CRC in the cecum, transverse and the descending/sigmoid colon. Majority of the patients (62.9%) with early-onset CRC had Stage III and IV disease upon diagnosis. Patients with early-onset CRC are less likely to harbor BRAF mutations (1.3 % vs 12.3%, p-value: 0.005). Microsatellite instability (MSI) is more common in patients with early-onset CRC with a trend towards statistical significance (23.9% vs 15%, p-value: 0.08).

Conclusion: There are unique demographic and clinical characteristics in patients with early-onset CRC. In addition, our study shows that molecular differences exist in patients who develop early-onset CRC. Further large studies are needed to validate our findings and to help identify characteristics unique to early-onset CRC so targeted efforts can be made to improve CRC screening in the higher-risk populations.
Targeting Notch signaling in glioblastoma cancer stem cells through modulation of Connexin43 function.

Authors: Michael Lunski, James W. Smyth, Jennifer Vaughn, Zhi Sheng, Robert G. Gourdie, Benjamin Purow, Samy Lamouille.

Introduction: Glioblastoma (GBM) is a malignant disease and even with the most current treatment regimens, which include a combination of chemotherapy (temozolomide), surgical resection and radiation, has a very poor prognosis. Though temozolomide (TMZ) initially works, its efficacy gradually decline. One hypothesis for why TMZ therapy ultimately fails is that the cells left behind after treatment, cancer stem cells (CSCs), have the ability to become resistant or may be unaffected, due to their quiescent state. For this reason, CSCs should be an area of focus. We know cellular mechanisms that are essential for cell stability. Notch is a highly conserved, signaling mechanism in most eukaryotic cells. It’s integral in neuronal development, proliferation and differentiation. Alteration of Notch and effect on cancer stem cell survival has not been studied extensively to date. In previous research, it has been shown that CSCs growth and stability can be interfered with by alteration of connexin 43 (Cx43), a gap junction protein, which interacts with microtubules. Recent studies, including our research, have shown that increased levels of Cx43 correlate with TMZ resistance in GBM cells and inversely correlated with GBM patient survival. Development of a novel Cx43 mimetic peptide, juxtamembrane 2 (JM2), incorporates the microtubules binding site on Cx43. JM2 has been shown to alter Cx43 activity. More importantly it has been discovered that there is a relationship between Cx43 and Notch, though the intricacies of this relationship have yet to be established. Our current research aims at dissecting the molecular mechanisms of Cx43 functions on Notch signaling in GSCs using this Cx43 mimetic peptide. In conclusion, we have identified a novel therapeutic opportunity to decrease the tumorigenic potential of these cells through altering Cx43 activity and Notch signaling to target chemoresistant GSCs in GBM treatment.

Methods: Human glioblastoma CSCs were obtained from tissue samples. Cells were cultured and plated into 6 well trays. Control cells were studied against treated cells, with JM2 peptide. Cells were then collected, and protein levels were assayed. Western blot was run with antibodies for Notch and Cx43 to assess for relationship. Coimmunoprecipitation was also performed. MG132, a proteasome inhibitor, was also used to assess for any change results.

Results: Initial results show a decrease in Notch with addition of the JM2 peptide, leading to further evidence of a relationship between Cx and Notch. Cell survival was clearly affected with addition of the peptide when looking under light microscopy as well.

Conclusion: Manipulation of Cx43 with JM2 confirms a relationship with Notch and decreased levels were associated with decrease in CSC survival. Notch targeting is likely an area that requires further research and may be an additional mechanism of therapy for a disease that desperately needs improved survival outcomes as well as a cure.
Reflexive to Reflective Lab Ordering - A Guideline Based Approach

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Introduction: One-third of all diagnostic testing is unnecessary, and approximately the same number of tests that could be useful are not being ordered (1). In 2012, an estimated $700 billion was wasted in American healthcare; overuse was identified as a large component of that waste ($280 billion) (2). Comparison of providers has been used to reduce diagnostic expenses among primary care providers. By educating primary care providers at the Salem VAMC on current guidelines through a multimodal approach and giving monthly progress reports, we aimed to reduce unwarranted lab ordering and ultimately the mean lab cost per unique patient seen by each provider.

Methods: After 6 months of gathering baseline lab ordering data for primary care providers at the Salem VAMC, a 6 month intervention period including education on guideline-based lab ordering and personal feedback was provided. Screening guidelines were first presented at Grand Rounds. A summary of guidelines from major societies was given to providers (attendings, mid-level providers and internal medicine residents). Providers were sent monthly comparison tables of their lab orders, including the mean cost per unique patient seen (lab reagent cost only, not personnel or other costs). Focused feedback during the intervention period was given to providers based upon their cost per unique patient seen or on opportunities identified from the actual lab studies ordered. Feedback also focused on avoidance of ordering labs in advance of patient visits. A subsequent 6 month follow-up period of gathering lab ordering data occurred; this was timed to occur during the same time of a calendar year as the baseline data was gathered.

Results: During the baseline period, the average cost per unique patient seen each month was $52.15. This progressively decreased during the intervention period. During the follow-up period, the average cost per unique patient decreased to $26.79. This translated into an annual savings of $385,000 (lab reagent alone). Through continued provision of lab ordering data and provider feedback, this cost reduction has been sustained (October, 2018 cost per unique of $24.69).

Conclusion: Over-ordering of diagnostic tests is a multi-million dollar problem across healthcare, with the VHA being no exception. Our primary goal was to decrease lab costs per unique patient for each provider without compromising the quality of care provided. After the intervention period of 6 months, cost per unique patient as well as total costs were dramatically reduced and then sustained with continued feedback. Similar results have been seen in regards to inappropriate antibiotic prescribing in VHA emergency departments (2). In the future, similar interventions could be implemented across specialty clinics as well as with inpatient providers, potentially further decreasing spending across the VHA medical system.

References

Prevalence and Indicators of Self-Reported Cognitive Dysfunction in Older Adults with Newly-Diagnosed gastrointestinal (GI) malignancies – results from the Cancer and Aging Resilience Evaluation (CARE) Study

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Introduction: Cognitive dysfunction is a poorly described phenomena in older adults with GI cancers. The purpose of this study was to quantify the prevalence of patient-reported cognitive impairment in older adults with GI cancers and identify baseline determinants associated with cognitive dysfunction.

Methods: This analysis draws from the CARE Study and includes patients aged ≥60y with a diagnosis of GI malignancy. Patients underwent a patient-reported Geriatric Assessment (GA) (CARE survey). Cognitive dysfunction was measured via the Patient-Reported Outcomes Measurement Information System (PROMIS®) Short Form 4a Cognitive Function survey. Descriptive statistics were used to examine the prevalence of cognitive dysfunction at baseline. Scores were dichotomized into normal/mild impairment (scores of 12-20) and moderate/severe impairment (scores of 4-11). Bivariate associations between demographic, clinical, and GA domains were tested to identify potential indicators of moderate/severe cognitive dysfunction.

Results: 159 older adults with newly diagnosed GI malignancy completed the CARE survey. Mean age of participants 69.6 ± 7.2 and 59.7% were male. Most common cancers included colon cancer (24.5%), pancreatic cancer (22.6%), and rectal cancer (12.6%). 96.2% of participants endorsed some level of cognitive dysfunction with their overall mean PROMIS-SF4a raw score 7.5±4. Nearly half of the participants endorsed severe dysfunction (47.2%), 35.2% of participants reported moderate dysfunction, while 13.8% of patients suffered mild symptoms. Cognitive impairment was negatively correlated with higher social activity limitations (ρ= -0.414, p<0.01) and higher PROMIS 10-item Global Physical and Mental Health scores (ρ= -0.405, p<0.01 and ρ= -0.495, p<0.01, respectively). Dichotomized cognitive dysfunction (none/mild vs moderate/severe) was also associated with the presence of Activities of Daily Living (ADL) impairment (36.7% vs 63.3%, p=0.01), instrumental ADL impairment (IADL) (27.7% vs 72.3%, p<0.01) and performance status ≥2 (37.7% vs 62.3%, p<0.01). The presence of hearing impairment (39.0% vs 61.0%, p<0.01), anxiety (51.5% vs 48.5%, p<0.01), and depression (71.4% vs 28.6%, p<0.01) was also associated with cognitive dysfunction. Lastly, no significant association was found with type or stage of cancer (p= 0.68 and p= 0.46, respectively).

Conclusion: We find a high prevalence of self-endorsed cognitive dysfunction in older adults with newly-diagnosed GI malignancies. These problems are associated with increased social activity limitations, ADL/IADL impairments, and mental health issues. Future longitudinal assessments of cognition after patients have received chemotherapy are planned to identify reliable predictors of cognitive dysfunction, and to facilitate the development of potential interventions.
How Trainees Finance their Medical Education: Implications of Higher Education Act Reform

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Introduction: Public Service Loan Forgiveness (PSLF) is one of many federal student loan forgiveness programs currently available for recipients of federal student loans. Medical education debt has continued to expand at a rapid pace in the past decade since PSLF was created. Recently proposed changes to the Higher Education Act (HEA) via the Promoting Real Opportunity, Success, and Prosperity through Education Reform (PROSPER) and Aim Higher Acts (AHA) would substantially change how future medical trainees finance and repay their medical education debt. To date, no one has directly assessed how growing medical education indebtedness impacts how residents are utilizing PSLF and other repayment tools as a mechanism of repaying education debt.

Methods: An IRB-approved anonymous survey was disseminated to all ACGME-accredited Internal Medicine residencies through direct e-mail contact of Program Directors. This instrument assessed resident personal loan burden, debt-associated stress, repayment plan, and potential use of PSLF.

Results: Data was obtained from 403 unique respondents at 12 residency programs, with a response rate of 69.6%. 80.2% reported indebtedness with a median value of $225,000. Education debt was reported to be a significant source of stress in 73.6% of these respondents. A majority (90.9%) reported a strategy for debt repayment, with Income-Driven Repayment and Standard Repayment being the most utilized methods at 77.1% and 15.6% respectively. Private loan use and loan forbearance was reported by 34.1% and 20.5%, respectively. While the majority of trainees were familiar with PSLF, only 40.6% report participation.

Respondents with the highest quartile of debt were more likely to have high levels of stress (OR 5.94, p<.0010) than those in the bottom quartile. Those without debt were more likely to have low levels of stress than those in the bottom quartile of debt (OR 10.44, p<.0001). Residents with higher debt burden were more likely to utilize PSLF (OR 3.269, p=.0241), while those with less debt were less likely to utilize (OR 0.287, p=.0052). Revised Pay As You Earn is utilized more frequently by those with higher levels of debt, and those with higher debt burdens are less likely to utilize the Standard Repayment option (OR 0.139, p=.0118). Higher levels of debt were associated with loan forbearance (OR 2.14, p=0.0283).

Conclusion: Graduate indebtedness is an influential variable affecting new physicians. Residents with higher debt burden have more stress and are more likely to utilize PSLF and enter loan forbearance. With three-quarters of medical residents utilizing income-based repayment plans and high levels of PSLF enrollment, future medical trainees are vulnerable to proposed public education policy changes that would eliminate or curtail these programs. Academic institutions, national medical organizations, and policy makers must scrutinize education cost, methods of financing, and repayment models as part of generating effective policy that enables future physicians to meet the health care needs of the United States.
Regional Differences in Epidemiology and Outcomes of Heart Failure Admissions Across the United States

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Introduction: Periodic surveillance of geographical variations in cardiovascular health is important to achieve the goal of reducing regional disparities in healthcare delivery. We aimed to study differences in epidemiology and outcomes of heart failure admissions by geographic regions in the United States.

Methods: We assessed the hypothesis that there exist differences in the outcomes of heart failure admissions based on geographic region. The National Inpatient Sample database for the year 2016 was queried. Adult patients admitted with a principal diagnosis of heart failure were identified using validated ICD-10 codes. Comparisons were made between four regions - Northeast, Midwest, South and West. Baseline characteristics of heart failure admissions were identified. The main outcomes of interest were inpatient mortality, length of stay and hospital charges. Statistical analysis was performed using STATA.

Results: A total of 807,764 hospitalizations with a principal diagnosis of heart failure were identified. Of these, 153,233 (18.97%) were in the Northeast; 184,090 (22.79%) in the Midwest; 331,506 (41.04%) in the South; and 138,935 (17.20%) in the West. The mean age, gender distribution and other baseline characteristics were similar between the regions. There was a small difference in the mortality rates between regions (highest in the West at 3% and lowest in the South at 2.66%, p=0.03). The length of hospital stay also differed between regions - longest in the Northeast at 5.66 ± 0.09 days; 5.29 ± 0.05 days in the South; 4.94 ± 0.06 days in the Midwest; and shortest in the West at 4.91 ± 0.07 days (p<0.001). A significant difference was observed in the total hospital charges per hospitalization - as expensive as $64,901 in the West; $52,289 in the Northeast; $44,886 in the South; and only $37,070 in the Midwest (p<0.001). The differences in all outcomes persisted after adjusting for variables like age, gender, race and co-morbid conditions.

Conclusion: Our study demonstrates the existence of regional differences in the costs and outcomes of healthcare delivery to heart failure patients. Further research is needed to explore the reasons for these differences.
The Impact of Benralizumab on Asthma Control, Asthma-related Quality of Life, and Lung Function in Patients with Poorly Controlled, Eosinophilic Asthma: A Systematic Review and Meta-analysis

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Introduction: Benralizumab is a monoclonal antibody to the alpha subunit of the IL-5 receptor used in the management of severe, eosinophilic asthma. While it has been shown to significantly reduce asthma exacerbation rates in several RCTs, its impact on subjective asthma control, asthma-related quality of life, and lung function remains less clear. The purpose of this meta-analysis is to analyze the combined effect of Benralizumab on Asthma Control Questionnaire (ACQ) scores, Asthma Quality of Life Questionnaire (AQLQ) scores, and pre-bronchodilator (pre-BD) FEV1 values in severe asthmatics with eosinophilia.

Methods: A comprehensive search of selected databases was performed to include randomized, phase 3 placebo-controlled clinical trials which compared the impact of Benralizumab on ACQ6 scores, AQLQ scores, and pre-BD FEV1 values in severe asthmatics with eosinophilia. Random effect models were produced to compare the combined effect of Benralizumab treatment in comparison to placebo.

Results: Overall, Benralizumab treatment in asthmatic patients with eosinophilia resulted in significantly improved ACQ6 scores, AQLQ scores, and pre-BD FEV1 values in comparison to placebo (Mean Difference: -0.24, 95%CI: -0.32,-0.16, p-value:<0.00001); (Mean Difference: 0.23, 95%CI: 0.14,0.32, p-value: <0.00001); (Mean Difference: 0.11, 95% CI 0.08,0.15, p-value: <0.00004) respectively.

Conclusion: Our meta-analysis demonstrates that treatment with Benralizumab in patients with severe asthma associated with eosinophilia significantly improves asthma control, asthma-related quality of life, and lung function. We believe these findings can provide evidenced-based recommendations for the use of Benralizumab in asthmatic patients with eosinophilia.
Managing Sepsis in Cancer Patients: A Retrospective Review of Oncologic Patients Presenting to Santa Clara Valley Medical Center

Authors: Charles Chu, MD; Ian Borrison, DO; Olivia Lee, MD

Introduction: Current sepsis guidelines are generalized towards the population regardless of underlying comorbidities. Few studies have investigated the management of sepsis in subgroup populations. Several studies have documented worse outcomes in sepsis in patients with cancer, however these studies did not address the difference in management between the two groups nor did they describe the physiologic features that may account for the worse outcomes. The advent of sepsis-3 introduced new sepsis definitions and introduced the quick sepsis related organ failure assessment (qSOFA) score to predict mortality in septic patients. The use of qSOFA in oncologic patients has not been validated as a marker for sepsis severity. Our primary objective is therefore to compare the clinical features of septic oncologic patients with that of the general population and to compare the efficacy of current sepsis management for oncologic patients. Our secondary objective is to validate the use of qSOFA as a marker of sepsis severity in a multicultural oncologic population.

Methods: We retrospectively reviewed 180 septic oncologic patients and 180 non-oncologic septic patients between July 2015 to September 2017. The primary outcome was mortality, length of stay, hospice, and disposition to nursing facilities. Additionally, we compared markers of sepsis severity including vital signs, lactic acid levels, and qSOFA scores. Finally compliance with sepsis core measures including volume of fluid resuscitation, compliance with lactic acid bundles and time to antibiotics were also compared.

Results: The cancer cohort had a lower systolic blood pressure (93 vs 102), a higher heart rate (120 vs 111) higher respiratory rates (27 vs 25). There was no difference between temperature, white blood cell count or creatinine. Average qSOFA scores were higher in the oncologic population at 1.6 compared to 1.2. The cancer cohort had a higher overall mortality at 16% compared to 6% in the non-cancer cohort, a longer length of stay at 13.9 days compared to 7.26 days, and more transitions to hospice at 9% compared to 2%. Lactic acid levels were also higher at 3 compared to 2.5. Both populations had 30% of patients who received 30cc/kg of crystalloid. Initial volume of crystalloid was 1.2L compared to 0.8L. There was no difference in time to antibiotics at 270 minutes.

Conclusion: Septic oncologic patients present with worse sepsis physiology and have higher rates of mortality, longer lengths of stay and transition to hospice with comparable rates of compliance with sepsis measures. It is unclear if current guidelines adequately treat septic oncologic patients. Our study shows no difference in time to antibiotics and similar rates of compliance with fluid resuscitation despite worse outcomes. Further studies would be needed to see if oncologic patients would benefit from different guidelines for sepsis management such as earlier antibiotics or a more liberal fluid strategy. qSOFA scores were higher in the oncologic population which validates qSOFA as a reliable predictor of mortality in this population.

References

How vital are vital sign checks? A quality improvement project to reduce unnecessary nighttime vital signs monitoring

Authors: Kelly Lo MD, Justin Yu, MD, Yoyo Jiang, MD, Ingeborg Schafhalter-Zoppoth, MD, Department of Internal Medicine, California Pacific Medical Center, San Francisco, CA

Introduction: Vital signs are important clinical markers, though optimal monitoring frequency is unclear. Hospitalized patients have regular vital signs checks regardless of clinical stability which can cause sleep interruption leading to increased use of sleep aids, delirium, and length of stay. At our hospital, vitals are checked every 4 hours, typically at: 0400, 0800, 1200, 1600, 2000, 0000. This default option is not routinely modified or reviewed by providers. In this project, we aim to reduce the number of nighttime vitals checks by 20% and to decrease number of sleep aids by 10%.

Methods: We used A3 thinking and PDSA cycling. In PDSA cycle 1, patients admitted to a resident team were monitored for 4 weeks. Number of nighttime vital checks between 2200 and 0400 were recorded. The first two weeks were used to establish current state (Pre) and the second 2 weeks were used to implement the following experiment (Post). The Modified Early Warning Score (MEWS) was used to calculate decompensation risk. Patients with MEWS <4 had vitals checks changed in EHR to “every 4 hours while awake” after discussion with primary team. In PDSA cycle 2, patients on 2 medicine teams were monitored for 1 week. Patient with MEWS <4 had vital checks changed to “every 4 hours while awake” using opt-out method starting on hospital day (HD) 2 and daily use of sleep aids was recorded.

Results: In PDSA cycle 1, the resident team had 92 patients over the 4-week period, with 48 in Pre and 44 in Post. There was 25%, 36%, and 38% reduction in vitals checks on HD1, HD4, and HD5. There was no statistically significant difference in the average MEWS between the two groups. One patient with a MEWS <4 decompensated overnight. Average length of stay was 6.6 in pre and 4.7 in post group. In PDSA cycle 2, 27.3% patients used sleep aids including melatonin, lorazepam and trazodone on HD1 compared to 15.8%, 16.6%, and 16.7% on HD 4, 5, and 6. Average length of stay 4.4 days.

Conclusion: In conclusion, modifying orders to vital signs ‘every 4 hours while awake’ resulted in >20% decrease in vital checks. It also led to a >10% reduction in patients using sleep aids. This QI project demonstrated that MEWS can help identify patients who may benefit from reduced vitals checks. Limiting vitals checks lead to decreased length of stay and sleep aideuse, though it is unclear if the intervention alone impacted this. Limitations of our experiment include lack of subjective patient survey data and lack of balancing measures. Future experiments include collaboration and education of care providers as patients still received vitals checks despite orders being changed and data acquisition on sleep quality and delirium.

References

Skin in the Game – A Multimodal Approach to Improving Resident Education for Skin of Color Dermatology

Authors: Shankar N. Mundluru, MD, Nirmala Ramalingam, MPP, Patrick McCleskey, MD, H. Nicole Tran, MD, PhD

Introduction: Diagnosis and treatment of dermatologic conditions play critical roles in disease prevention. However, previous studies demonstrate that dermatology education in medicine residency is underemphasized, and education for skin of color dermatology is especially limited. We sought to address this knowledge gap at the Kaiser Oakland Internal Medicine Residency.

Methods: We performed a root cause analysis of current and gap conditions, and we implemented three education sessions based on this analysis.

Results: We first highlighted the importance of this topic during a traditional lecture, a hospital wide grand rounds presentation. We discussed the epidemiology of skin of color diseases and highlighted their lack of representation in textbooks, conferences, and residency education. We also gave information about important resources, such as websites, articles, and textbooks, and we encouraged a system wide effort to augment any dermatologic images with darker skin. We then led a two-hour flipped classroom during which residents taught each other about common skin of color conditions, and we taught residents tools to take better pictures of conditions on dark skin. Finally, we developed two games for residents to play during a 30-minute session. “Skin Matchmaker” is a game through which residents were given 20 images of dermatologic conditions: 10 images of conditions on dark skin and 10 images of identical conditions on lighter skin. They matched identical conditions and described similarities and differences in appearance and treatment considerations. “Guess Who? The Derm Term Version” includes a group of 4 residents who were given 10 images of conditions on dark skin. Three residents were given one image of the 10 to provide clues, such as visual descriptions, treatment, and epidemiology, to the fourth resident who had to guess the correct image.

Conclusion: The traditional lecture was best used to introduce simple and easily understood concepts, and the flipped classroom session allowed residents to absorb important, clinically applicable information in an interactive, group setting. The gaming session best engaged individual learners in visual learning and pattern recognition, and it allowed residents to learn at their own paces. The interactive, visual learning increased motivation to learn and permitted transmission and improved comprehension of complex topics. Our multimodal approach could be used as a model for early intervention in addressing residents’ deficits regarding skin of color dermatology.

All the sessions were well received by residents, as over 85% rated the sessions as very good or excellent. Based on feedback, we will implement an online interactive module in the spring that includes the above games in addition to information on skin of color dermatology. Once implemented, we will send a post intervention survey assessing residents’ knowledge and comfort of ethnic dermatology, and we will utilize these results to further refine these sessions.
Social Vulnerability Index Correlates with Rate of Asthma Related Emergency Department Visit and Hospitalization

Authors: Sandeep S Nayak, MD MS., Dan Pham, MD., Laren Tan, MD

Introduction: Prevalence, emergency department (ED) visits, and hospitalizations due to asthma vary from one county to other and are higher among women than men. Biological, genetic, and immunological factors contributing to this gender difference has been extensively studied. Socioeconomic factors influencing this, however, have not been well studied. Social Vulnerability Index (SVI) is a surrogate for population-based measures of socioeconomic well-being. SVI with the values ranging from 0-15, is a sum of values of four related themes, namely (1) Socioeconomic status (2) Household Composition and Disability (3) Minority Status and Language and (4) Housing and Transportation. Objective of this study to evaluate if Social Vulnerability Index (SVI) correlates with ED visits and hospitalization in adult asthmatics.

Methods: This is a cross sectional study involving secondary analysis of county level data from 2014 in the United States (USA). SVI data was obtained by CDC. Data on age-adjusted rate of hospitalization and ED visits for asthma (per 10,000 population) was obtained by CDC’s National Asthma Control Program. Simple linear regression model results and corresponding Pearson correlation coefficients were calculated for overall SVI and subgroup.

Results: Out of 3142 counties in USA, data on hospitalization was available from 1383 counties for females and 1360 for males. Data on ED visit was available from 1050 counties for males and females. In females, for every 1 unit increase in SVI, there was 0.599 increase in the rate of hospitalization and 3.729 increase in the rate of ED visits (p < 0.0001). In males, for every 1 unit increase in SVI, there was about a 0.378 increase in the rate of hospitalization and 2.813 increase in the rate of ED visits (p < 0.0001). Analysis of individual themes demonstrated strongest correlation with theme 3. For every 1 unit increase in theme 3, there was about a 1.295 increase in the rate of hospitalization and 8.831 increase in the rate of ED visits (p < 0.0001) for females. In males, for every 1 unit increase in Theme 3, there was about a 1.703 increase in the rate of hospitalization and 10.677 increase in the rate of ED visits (p < 0.0001).

Conclusion: There is a significant association between the social vulnerability index and both the rate of hospitalization and the number of emergency department visits, regardless of gender. Also, social vulnerability has stronger correlation with females than males which may explain multifaceted social characteristics of each county influencing on gender disparity in Asthma. In addition, minority status and language seems to be having strongest correlation with ED visits and hospitalization.
CALIFORNIA RESEARCH POSTER FINALIST - ERICA DUH, MD

Achieving an Optical Artificial Intelligence (AI) System for Real-Time Diagnosis and Resection of Colon Polyps


Introduction: Colorectal cancer (CRC) is the second leading cause of cancer deaths in the United States. 70-90% of CRC are preventable with removal of polyps, however, many polyps are deemed “diminutive” (less than 5mm in size and are rarely malignant). Pathology fees for evaluation of these polyps contribute towards the $1 billion annual cost of colonoscopies alone. A real-time method of optical diagnosis of these polyps has the potential for enormous cost-savings. We developed an artificial intelligence (AI) optical biopsy system that meets the Preservation and Incorporation of Valuable Endoscopic Innovations (PIVI) "Resect and Discard" guideline set forth by The American Society for Gastrointestinal Endoscopy’s (ASGE). It requires a > 90% negative predictive value (NPV) for diminutive adenomas and > 90% concordance in assignment of surveillance intervals when compared with decisions based on pathology. Convolutional neural networks (CNN) have the potential to predict polyp pathology and meet PIVI guidelines independently of operator or scope manufacturer.

Methods: Using Qualoscopy — a UCI Colonoscopy Quality Data base made up of images pulled from over 10,000 procedures we pre-trained a CNN built on Tensorflow. Images of adenomas and polyps of varying locations, size, and light source (white light [WL] or narrow band imaging [NBI]) were partitioned into 5 equal-sized subsamples for 5-fold cross validation with training (80%) and validation (20%). An Adam optimizer generates a probability between 0-0.5 (serrated) and 0.5-1 (adenoma). Surveillance intervals were calculated based on US Multi-Society Task Force guidelines, comparing OP versus true pathology (TP).

Results: The NPV for adenomas was 92% (WL) and 93% (NBI) for polyps that were found throughout the colon. Surveillance interval concordance between OP and TP for screening and surveillance cases was 93% and 96%, respectively. Among diminutive polyps (< 6 mm) throughout the colon, NPV for adenomas was 91% (WL) and 92% (NBI). Surveillance concordance was 93% and 96% for screening and surveillance cases, respectively. When looking at diminutive polyps in the left colon, NPV improved to 97% (WL) and 95% (NBI). The model processes more than 90 frames per second and can be rendered in real-time during colonoscopy using a conventional desktop and graphics processing unit.

Conclusion: We demonstrate feasibility of an optical AI biopsy system that provides operator-independent and real-time feedback during colonoscopy. The optical AI biopsy system provides a ≥ 90% agreement in assignment of post-polypectomy surveillance intervals compared with decisions based on pathology as outlined by the PIVI “Resect and Discard” guidelines. This system also provides immediate data in regards to adenoma detection and surveillance recommendation at the time of colonoscopy. We hope to validate our system through blinded multi-center studies utilizing multiple scope manufacturers in the near future.

References

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What are the Social Determinants of Deciding Resuscitation Status ("DNR" vs. "Full Code") in Critically Ill Patients > 70 years?

Authors: Julius Musenze, DO (PGY3) and Christina Vu, DO (PGY2), Mentor: Dr. Learned Gonzales, MD (Pulm/Crit)

Introduction: We set out to investigate whether racial identification, religion, education level, occupation, income, means by which a code decision is reached (individual vs. group), relationship of decision maker to the patient, and the age of patient or age of decision maker, influenced resuscitation status. Prior study by Jorge, et al (2000) was limited to patients with acute myocardial infarction but it concluded that DNR assignment was inversely associated with black race and positively associated with age, probability of death, cognitive impairment, and poor nutritional status. Another study by Dong, et al (2014) in 77,329 hospitalized septic patients concluded that smaller hospital size, absence of teaching programs, greater patient age, female gender, White race, medical comorbidities, Medicare payer status and admission from a skilled nursing facility were all significantly associated with an early DNR designation.

Methods: n=77: We interviewed all patients 70 years and older admitted at Desert Regional Medical Center on Saturday August 4th 2018. This “entire-hospital-on-a-single-day” approach was designed to eliminate selection and sampling biases. We explained the research study and made it clear that agreeing or declining to participate had no bearing on the level of care they received. Interested participants (52/77) were consented and given questionnaire in their respective first language (English vs. Spanish). 25/77 were excluded due to: “patient confused or sedated” (3/25), “patient off the floor for procedure” (9/25), “patient sleeping” (1/25), “patient deceased” (1/25), “patient discharged” (1/25) and “patient/family declined” (10/25).

Results

Positive Correlation:

1. Income: Higher Income individual were more likely to be “DNR”: (t-value = -2.774; p=0.008 < 0.05)
2. Religion: Catholics/Christians were overwhelmingly Full Code while Atheists opted for DNR: (χ2 = 17.814; p = .007 < 0.05)
3. Education: “Some schooling/Some College” preferred Full Code while “University grad/masters” opted for DNR: (χ2 = 6.842; p = .033 < 0.05)

No Correlation:

Place of Birth (χ2 = .039; p = .843 > 0.05), Race (χ2 = .365; p = .947 > 0.05), Age (t = -1.385, p = .185 > 0.05), Gender (χ2 = .620; p = .431 > 0.05), Individual Vs. “Group Think” (χ2 = .277; p = .599 > 0.05) and Knowing the diagnosis (a way to assess mental competence) (χ2 = .056; p = .813 > 0.05) had no bearing on code designation.

Conclusion: We concluded that Income level (Higher income=DNR), Religious beliefs (Catholics=Full Code, Atheists=DNR) and Education level (Some education=Full Code, University Graduate=DNR) influenced a patients’ code designation. DNR or Full Code assignment was not associated with birth place, race, patient’s age, patient’s gender, decision maker, number of decision makers or knowledge or diagnosis. As clinicians, understanding these social determinants will help us better understand why patients and their families choose one over the other and thus facilitate better communicators when it comes to the matter.
References


CENTRAL AMERICA RESEARCH POSTER FINALIST - MIRIAM MARIA GARCIA FALLAS, MD

"THE IMPACT ON HEALTH IN PATIENTS OF THE CARDIAC REHABILITATION PROGRAM IN THE PERIOD 2013-2015 IN THE CENARE"

Authors: Dr. Miriam Garcia Fallas Resident of Internal Medicine HSJD Costa Rica

Introduction: Mortality from heart attacks from 2000 to 2008 shows a decrease significant in relation to the comparison between rates. Which implies that more patients survive a coronary event and require a process of cardiac rehabilitation that allows to diminish its morbidity and mortality and join the economically active population. The objective of the study was to analyze the impact on health in patients who participated in a program of cardiac rehabilitation in the period 2013-2015 at the National Center of Rehabilitation

Methods: A retrospective observational study was carried out. The population studied were patients who completed Phase II with a diagnosis of ischemic heart disease. Analysis of descriptive and inferential statistics of the data was carried out.

Results: A total of 460 records were reviewed. A sample of 272 patients was obtained, with a total of 75.9% male and 24.1% female. The average age was 64.7 years. In the metabolic variables, the behavior was the decrease in concentrations of LDL cholesterol, total cholesterol and triglycerides, without reaching the value goals. HDL levels increased in all three periods. The change in METS achieved in the stress tests was statistically significant. There was an average increase of 2.73 METS during the 10 weeks of training

Conclusion: The decrease in the values ??of the lipid profile had statistical significance but without reaching the target value, we could infer that these goals will be achieved during Phase III.

The values ??of arterial pressure had a tendency to decrease, as the benefit in controlling blood pressure and the vasodilation arteriolar response as a reflection of the improvement of the maximum consumption of oxygen mediated by the decrease in endothelial dysfunction.

Physical training improves the frequency of recovery. Patients with persistently high frequencies after physical exercise have a higher cardiac mortality. In the population studied, the response in cardiac recovery frequency remained below 10 beats. This is important, due to its clinical application as a therapeutic goal and is one of the parameters that predict mortality in these patients. The functional capacity that was measured increased significantly. This effect is the result of changes in the cardiovascular system and the improvement in muscle strength, the change in metabolism of skeletal muscle fibers that improve with aerobic exercise, thus, managing to increase tolerance to workloads and locally improve blood circulation.

The improvement in functional capacity is the most relevant contribution, since it may be associated with a decrease in overall mortality, given that for each increase of a METS the overall mortality is reduced by 8 to 14%.
Effects of medical cannabis and cannabinoids: Living OVerview of the Evidence (L-OVE)

Authors: Oscar Corsi, Mariaignacia Morales, Francisco Allende, Diego Lobos, Carolina Nuñez, Rami Guinguis, Tania Contreras, Matías Rocco, José Peña, Pedro Pérez, Carlos Juri, Eugenio Maul, Pedro Ortiz, Macarena Morel, Cynthia Zavala, Gonzalo Bravo, Gabriel Rada

Introduction: The existing clinical research about the therapeutic effects of cannabis and cannabinoids is controversial. New studies on this topic are published at a fast rate, and their number grows larger by the day. Keeping pace with the evidence has become difficult and dozens of systematic reviews are published each year trying to synthesize this body of evidence.

LOV-E is an Epistemonikos Foundation’s project which organizes all the updated evidence relevant for making decisions related to a specific healthcare topic.

Our objective is to assess the therapeutic effects of cannabis, cannabis-derived products and synthetic cannabinoids for multiple health conditions using this new strategy.

Methods: We conducted a search in Epistemonikos, the world’s largest systematic review database. Epistemonikos is maintained by screening multiple information sources to identify systematic reviews and their included primary studies, including Cochrane Database of Systematic Reviews, Pubmed/MEDLINE, EMBASE, LILACS, DARE, among others. At least two reviewers independently screened titles and abstracts to identify relevant articles.

For some of the identified health conditions, we made an evidence matrix, a visual interface that compares all systematic reviews addressing a similar question. Then, the team extracted the relevant information from the systematic reviews, creating a Living FRiendly Summaries of the Body of Evidence using Epistemonikos (FRISBEE) which included all the body of evidence in a user-friendly format.

Results: There were 618 possibly relevant reviews of which 161 were selected. These include 603 primary studies (438 are randomized). By now, 15 FRISBEEs have been published. The conclusions fall into one of 3 categories:

a) Cannabis or its derivatives are not effective and are associated with frequent adverse effects (multiple sclerosis and anorexia nervosa).

b) Cannabis or its derivatives may produce a low benefit, but also produce frequent adverse effects that exceed the benefits (cannabis abuse disorder, chronic pain, and epilepsy).

c) It is not clear whether cannabis or its derivatives are effective or not, because the certainty of the evidence is very low, and they are associated with frequent adverse effects (fibromyalgia, insomnia, Tourette’s syndrome, and the management of nausea/vomiting induced by chemotherapy).

Conclusion: Through a novel method, we have synthesized a large volume of evidence on the therapeutic effects of cannabis and cannabinoids. The ongoing research is considerable and we hope to include all the health conditions for which the use of cannabis and cannabinoids has been proposed and to maintain the reviews continuously updated, since there may be new relevant information in the future.
Oral Vancomycin as an effective monotherapy for the treatment of primary sclerosing cholangitis

Authors: Tsay, Cynthia J., Lemos, Leta, Scudiere, Jennifer, Cox, Kenneth L., Davies, Yinka K.

Introduction: Primary sclerosing cholangitis (PSC) is a rare, autoimmune inflammatory disorder of the liver leading to destruction and inflammation of intra- and extra-hepatic bile ducts. Oral vancomycin has been reported to treat PSC and associated inflammatory bowel disease by its possible immune-modulating effects secondary to alteration of the gut microbiome in patients with PSC, however studies have reported changes in serum biochemistry without looking at histological changes.

Methods: In this prospective case series, we report 13 patients who received liver and large intestine biopsies at the time of diagnoses with PSC and after treatment with oral vancomycin at a single provider practice. Serum was also drawn at these two time-points and throughout treatment. Histology was interpreted by a single, blinded pathologist and graded on a pre-determined system aimed to allow comparison between the two time-points.

Results: The final analysis involved 13 patients, 23% were female, with an average age of 13.9 at the time of diagnosis and 15.7 at the end of treatment. Among the 12 lower gastrointestinal (GI) biopsies, 58% had improved histology after treatment. For the 13 liver biopsies, 69% demonstrated disease amelioration when compared to biopsies at the time of diagnosis (Figure 1). There was a statistically significant improvement in alkaline phosphatase (0.018), AST (p=0.003), ALT (p=0.002), and GGT (p=0.003). Using the W-value from the Wilcoxon-signed rank test, CRP and ESR were also found to be statistically significant (p<0.05).

Conclusion: Oral Vancomycin not only leads to biochemical improvements in liver transaminases in PSC but also histological and clinical improvement by halting disease progression as seen in pre- and post-treatment liver and lower GI biopsies.
Value Added Risk Assessment Using Coronary Artery Calcification from Non-Contrast Chest Computed Tomography (CT) Images

Authors: Pankil Desai, MD; Weichun Wu, MD; Lloyd Xiao, BS; Gregory DN. Pearson, MD, Ph.D.; Suzanne Rose, Ph.D.; David Hsi, MD

Introduction: Coronary artery calcification (CAC) from the non-contrast chest CT scans may provide insight to the patients’ coronary artery disease (CAD) risks and influence the early intervention process. Multi-Ethnic Study of Atherosclerosis studies revealed that CAC is a powerful risk predictor for CAD and is widely used in the national practice guidelines. The current clinical practice of reporting non-contrast chest CT does not include uniform assessment of coronary artery calcium deposits.

Methods: We sought to review available CT imaging data and retrospectively estimated the CAC burden in a general patient population over a 3-year period. The research protocol was reviewed and approved by the central research office in our hospital. We included 500 consecutive patients, 231 males and 269 females with an average age of 74 years old who had non-contrast chest CT at our hospital for any indication. Patients with intravenous contrast chest CT, prior coronary artery stent placement or CT angiogram were excluded. A scoring system (Weston Score) was used to assign a score for each major coronary vessel as follows: 0—no visually detected calcium; 1—a single high-density pixel; 3—calcium dense enough to create blooming artifact; and 2—calcium in between 1 and 3. The Weston scores were calculated by the sum of the score for each vessel (range 0–12). Mortality data were obtained from social security death index and electronic medical records.

Results: There were total of 56 deaths. The risk of death was related to the presence of coronary calcification, p=0.005; it was also related to calcification of any coronary artery, p<0.05. The severity of the calcification by the Weston Score, however, was not directly related to the risk of death, p=0.328. In 357 patients with CAC 41% were on statin drugs; 31% were on aspirin; and 22% were on both statin and aspirin.

Conclusion: In general population, CAC is related to risk of death and easily identified from non-contrast Chest CT images. Many patients can benefit from early treatment for CAD based on the clinical profile including CAC data. The presence, not the severity of CAC influencing mortality risks indicated the primary importance of visual recognition of CAC in the radiology report, while the semi quantitation of the calcium burden may help individualization of patient management. There might be potential cost savings of not obtaining dedicated CT calcium scoring in some patients if the prior non-contrast chest CT data was available for analysis. Our results also demonstrated relatively low utilization of aspirin and statin drugs for the primary prevention of CAD in this nonselective patient cohort. Clinicians and radiologists can maximize the value of non-contrast chest CT by reporting CAC. We can use this powerful data to identify patients with existing CAD and optimize medical management for patients with unrecognized CAD.

References

Helping with HELP (Hospital Elder Life Program) – A Needs Assessment, Program Implementation and Cost-Effective Analysis.

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Introduction: Delirium is a common, but preventable, problem that affects up to 60% of hospitalized older adults. Up to 40 percent of delirium cases are preventable (Inouye, Lancet, 2014). Delirium is associated with many negative outcomes, including falls, increased hospital length of stay (LOS), functional/cognitive decline, institutionalization and mortality. Delirium is also associated with significant healthcare costs. The Hospital Elder Life Program (HELP) is a comprehensive program of care based on award-winning clinical trial designed to reduce delirium and its complications in hospitalized older adults. The purpose of this study is to establish the needs assessment and cost-effective analysis to support the benefit of instituting the HELP program and demonstrating reduced rates of delirium, LOS, fall rates and readmission rates post HELP program implementation at a community level hospital.

Methods: A Cross-Sectional Analysis of de-identified data from Griffin Hospital Electronic Medical Records was conducted for fiscal year 2016 to determine the number of hospitalized elders affected by delirium. Various outcomes among those who met inclusion criteria “with delirium” were compared to those “without delirium”. The outcome measures consisted of determining the rate of delirium, length of stay, rate of hospital falls and rate of readmission. Subsequently, cost-effective analysis and implementation plan was also conducted using HELP Business Model and post intervention data was analyzed in the interim.

Results: Records were reviewed for the total of 7598 admissions at Griffin Hospital for fiscal year 2016. Among the target population, 25.8% (1956) met the inclusion criteria. Among those who met the inclusion criteria, 12.8% (248) were those that were 70 years and older, 11.04% (216) were those that had length of stay > 2 days. The average length of stay was 5.7 days for those “without delirium” which increased to 6.7 days for those with diagnosis of delirium. A 2-fold increased risk of falling was observed among those with delirium (prevalence ratio=2.0125). Using the Business model supplied by the pioneers of the HELP Program, a cost-effective analysis was conducted, which demonstrated, that this volunteer-based program will pay for itself after an initial implementation. HELP was implemented in February 2018 and by end of September 2018, a total of 198 patients were enrolled in the HELP Program. Interim Analysis of post intervention (HELP Program Implementation) data demonstrated extremely encouraging results with Average LOS among those with Delirium noted to be 4.7 post intervention compare to 5.7 pre intervention (P<0.05). The rate of new onset delirium was found to be 2.5% with fall rate being 0.5% post intervention compare to 0.87% pre intervention (p<0.05). Updated results will be presented at the ACP Meeting.

Conclusion: After conducting successful needs-assessment, cost-effectiveness analysis and program implementation, interim analysis of post HELP Program implementation data support the ability of the program to reduce hospital-acquired delirium rates, LOS, and fall rates.

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Rates of No-Shows in an Urban Primary Care Residency Clinic: Defining the Problem

Authors: Elizabeth Pines, DO, MS; Patrick Ho, DO; Spencer Hall, Olivia Gad, DO; Shannon Wilton, DO; Michaela Seigo, DO; Patty McGraw, RN, MS; Vishal Patel, MD, MBA; Cheryl Jackson, MD; Gretchen Rickards, MD, MPH

Introduction: No-show rates among primary care practices in the United States vary from 5% to 55%. Missed appointments have been associated with increased acute care utilization, reduced access for other patients, worse health outcomes, and decreased provider productivity. Continuity clinic is a required component of residency training and missed appointments translate into missed educational opportunities. Few studies have looked at the differences between no-shows among residents and attendings as well as internal medicine residents and family medicine residents in the context of primary care. The aim of this study was to examine the association between no-show rates at an urban internal medicine and family medicine residency continuity clinic based upon provider characteristics (i.e., specialty type and post-graduate year) and appointment type as well as the financial implications no-show appointments have on our clinic.

Methods: We performed a retrospective chart review of all scheduled appointments from July 1, 2016 to December 4, 2017 of adult patients at an urban internal medicine and family medicine residency continuity clinic. Data was obtained from a single large regional mid-Atlantic health care system. A no-show appointment was defined as an appointment where the patient failed to appear and did not cancel prior to the appointment time. Lost clinic revenue was calculated based on the following billing codes: 99204, 99213, 99214, 99243, 99396. Chi-square test was employed to analyze differences between internal medicine providers and family medicine providers.

Results: During the study period, there were 47,753 discrete visits. The overall no-show rate was 23.3% (n=11,137), which differed by provider type with internal medicine residents having the highest no-show rate (29.0%) when compared to family medicine residents (26.3%), internal medicine nurse practitioners (19.1%), and all attendings (17.9%) (p<0.01). No-show rate by post-graduate year was significantly different only among categorical internal medicine residents with the later post-graduate years having lower no-show rates (p < 0.0001). For both the internal medicine and family medicine practices, hospital discharge follow-up appointments had the highest no-show rate (34.7% and 38.5%, respectively) while pre-operative appointments had the lowest (9.1% and 17.6%, respectively). The clinic had an estimated $2,092,260 in lost revenue from no-show appointments during the 18-month study period.

Conclusion: This study identified significant differences in no-show rates between residents and attendings as well as internal medicine residents and family medicine residents. Hospital discharge follow-up appointments had the highest no-show rates among all residents. The next step of this project is to develop and implement a survey to assess patient barriers to attending appointments with emphasis on transportation, financial, and scheduling concerns. Subsequently, the chart review and survey data will be combined to develop targeted interventions to reduce no-show rates.

References

DELAWARE RESEARCH POSTER FINALIST - ELIZABETH M PINES, DO

Barriers to Attending Appointments in an Urban Internal Medicine Residency Clinic: A Survey Study

Authors: Elizabeth Pines, DO, MS; Patrick Ho, DO; Spencer Hall, Olivia Gad, DO; Shannon Wilton, DO; Michaela Seigo, DO; Esteban Mercado-Rodriguez, MD; Jeffrey Shuman, MD; Patty McGraw, RN, MS; Vishal Patel, MD, MBA; Cheryl Jackson, MD; Gretchen Rickards, MD, MPH

Introduction: No-show rates among primary care practices in the United States vary from 5% to 55%. Missed appointments have been associated with increased acute care utilization, reduced access for other patients, worse health outcomes, and decreased provider productivity. Patients cite many reasons for not attending appointments. However, few studies have looked at barriers of attendance among patients of a residency clinic. The objective of this study was to identify barriers to attending primary care visits among adult patients who “no-showed” at an urban residency clinic.

Methods: A nine-item closed-ended telephone survey of sequential patients who “no-showed” for primary care appointments from May 29, 2018 to June 29, 2018 was completed by trained research assistants over a three-month period. Questions focused on barriers related to money (e.g., could not afford transportation, could not afford time off from work), transportation, and scheduling (e.g., trouble getting time off from work, not being able to get an appointment soon enough). The survey included demographic questions, i.e., race, gender and insurance status. Other demographic data such as age and street address as well as appointment information was obtained from the medical record. Frequencies and means were used to describe patient characteristics and demographics. Chi-square test was used to compare characteristics of responders vs. non-responders.

Results: During the study period there were 422 no-show appointments, which represented 391 patients. Two hundred sixty patients were contacted and 72 (27.7%) completed the survey. Responders, when compared to non-responders, were more likely to be female (80% vs. 51.6%), black (79.2% vs. 64.9%), have Medicare (30.6% vs. 23.4%), and be a patient of an attending (30.6% vs. 17.0%). The most common (54.1%) reason provided for no-show was forgetting the appointment. Over 60% of respondents did not cite any financial, transportation, or scheduling barriers to attendance. For those who experienced barriers, issues with scheduling was most common (n=32, 44.4%) followed by transportation (n=23, 31.9%). However, when asked more specific barriers, transportation was ranked as the top three reasons for no-show. Furthermore, 98.6% of responders stated reminders of upcoming appointments would be helpful. Of those, 60% wanted a cell phone call and 55.6% wanted a text message while only 31.9% wanted a home phone call reminder for their appointments.

Conclusion: While most of our patients did not report experiencing barriers to attending primary care appointments, those that did mostly experience barriers to reliable and affordable transportation and receiving reminders in the desired form. While the responders and non-responders were very different, we identified to modifiable contributors to no-show rates at our urban internal medicine residency clinic, reminder system and transportation. The next step of this project is to develop and implement interventions to improve the reminder system and work with barriers to transportation through innovative health care delivery.

References
DELAWARE RESEARCH POSTER FINALIST - BRYAN A HAIMES, MD, MPH

Improving Call Experience Using KUMQUAT (Call Monitoring Quality Tool): 10 month follow up on tool use and efficacy

Authors: Bryan Haimes, MD, MPH; Kristen Isaac, MPH; Neil Gaskill, DO; Dhara Shah, DO; Brianna Buzzuro, MSN, RN; Victoria Varga, MSN, APRN; Mukarram Razvi, DO; Loretta Consiglio-Ward, MSN, RN

Introduction: Patient experience is at the forefront of improvement projects in the new value-based healthcare reimbursements to help improve the perceived value of healthcare.\(^1\) To improve patient experience, it is important to consider all interactions patients have with the healthcare system. A substandard experience at any point of contact can lead to poor patient compliance, adverse events, and/or worsened clinical outcomes. Patients need to experience good communication and ease of communication with healthcare professionals, easy access to care and information, good customer service, and good coordination of care.\(^2\) The Medical Group Call Center (MGCC) is a new concept at Christiana Care Health System that aims to improve patient-practice interactions to ultimately optimize patient outcomes. The MGCC currently takes calls for eight primary care and three specialty practices and is intimately involved in the aforementioned patient experience aspects of healthcare. At its inception, the MGCC had no tools to efficiently, effectively, and routinely monitor call quality. Patients have expressed significant frustration regarding poor experiences with the MGCC which has led to escalated calls and adverse clinical events. This project’s goal is to provide a tool to give feedback to call center staff to improve quality of call experiences.

Methods: MGCC supervisors were consulted to develop an internally validated Call Monitoring Quality Tool (KUMQUAT) using REDCap software to capture specific elements in 4 categories of call quality: greeting/ending, personal, computer skills, and procedure. Algorithms were developed to calculate scores within each category. Guided by Plan-Do-Study-Act (PDSA) framework, MGCC supervisors utilized KUMQUAT during a two-week trial period to monitor and evaluate patient service representative (PSR) calls, questions were changed and tool was altered based on feedback. The revised KUMQUAT has been implemented over the past 10 months.

Results: Prior to implementation, call quality and escalated call quantity were not routinely monitored by the MGCC. By completion of the initial two-week trial, 66 calls were monitored, 0 of which were escalated. Since the alterations of the KUMQUAT in mid-June, this tool has been used to monitor 123 calls per month (SD 30 calls). This has led to subjective and objective improvement in call quality, with KUMQUAT scores increasing from 64\(\%\) during the initial PDSA cycle up to 96.6\(\%\) over the past six months. The algorithms have been enhanced to provide automatic, customizable, weekly reports on call quality.

Conclusion: This project created a mechanism for Call Center supervisors to monitor call quality, and collect ongoing quality data that can be used to direct future PSR training and development. Since implementation of this tool 10 months ago, data demonstrate overall improvement in PSR call quality. The PDSA process facilitated the collection of objective feedback and capturing of real time call quality to improve interactions of patients with the healthcare system. This project allows continued improvement in the patient experience.

References


Use of Whole-Genome Sequencing to Guide a C. difficile Diagnostic Stewardship Program

Introduction: Hospital-onset C. difficile infection (HO-CDI) has been problematic at our hospital, with rates almost 50% greater than predicted. C. difficile whole-genome sequencing (WGS) data was used to define the transmission pattern, followed by a diagnostic stewardship intervention.

Methods: Isolates from CDI cases were sequenced for strain relatedness and epidemiologically analyzed using a single nucleotide polymorphism (SNP)-based approach. In June 2017, a diagnostic stewardship intervention began which included provider education and a weekday review of CDI orders placed after hospital day 3 for the following indications: >3 stools/24 hours, the absence of laxative administration, the presence of fever/leukocytosis or a history of inflammatory bowel disease. In Nov 2017, an EMR-based testing algorithm was introduced to supplement the review process. Orders not meeting testing criteria were discussed with the ordering provider, with a suggestion to cancel orders without appropriate indications.

Results: WGS assigned 36 isolates to 19 different multi-locus sequence types (ST), including 5 assigned to ST-1, a sequence that encompasses the ribotype 027 clade (Figure 1). SNP-based analysis indicated closely related, but non-identical strains, inconsistent with nosocomial transmission. 646 CDI orders were reviewed, of which 421 (65%) met criteria and 64 (15%) were positive. 225 (35%) of orders were recommended for cancellation. The HO-CDI rate decreased from 11.67/10k in the 5-month baseline period to 7.13/10k in the 9-month intervention period (p=0.0008) (Figure 2).

Figure 1

Dendogram of Isolates

<table>
<thead>
<tr>
<th>CD199-Ribotype-27</th>
<th>R20991-Ribotype-27</th>
</tr>
</thead>
<tbody>
<tr>
<td>17-083-00878</td>
<td>16-149-01063</td>
</tr>
<tr>
<td>16-141-12267</td>
<td>16-139-08888</td>
</tr>
<tr>
<td>17-367-13322</td>
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</tr>
<tr>
<td>16-342-04765</td>
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</tr>
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<td>M88-Ribotype-17</td>
</tr>
<tr>
<td>17-086-07665</td>
<td>17-086-07665</td>
</tr>
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<td>17-053-03232</td>
</tr>
<tr>
<td>17-051-06955</td>
<td>17-051-06955</td>
</tr>
</tbody>
</table>

Figure 1
Conclusion: WGS revealed that nosocomial transmission of *C. difficile* was an unlikely cause for our elevated CO-CDI rate. A diagnostic stewardship intervention which focused on identifying community-acquired infection and avoiding over-testing was associated with a sustained decrease in the HO-CDI rate which has persisted for 9 months.
DISTRICT OF COLUMBIA RESEARCH POSTER FINALIST - TRAN NGUYEN, MD

ANTIHYPERTENSIVE TREATMENT TO LOWER CARDIOVASCULAR RISK AMONG POST-MENOPAUSAL WOMEN ON ESTROGEN REPLACEMENT THERAPY

Authors: Tran Nguyen, MD; Talal Alzahrani, MD, MPH; Jannet Lewis, MD, FACC., Department of Medicine, The George Washington University, Washington, DC

Introduction: Hypertension remains the number-one “silent killer” in the United States due to its prominent causative connection with heart disease and stroke. Approximately one in every three women between the ages of 40 to 59, and two in every three women aged 60 and above in the United States have hypertension. For older women, estrogen supplementation is occasionally used to treat common menopausal symptoms, but increases the risk of cardiovascular events. Although the results of the original Antihypertensive and Lipid-Lowering Treatment to Prevent Heart Attack (ALLHAT) trial showed that hydrochlorothiazide (HCTZ) diuretics are superior to calcium-channel blockers and angiotensin-converting enzyme inhibitors in preventing 1 or more major forms of cardiovascular disease (CVD), there are several reasons why physicians may prefer to avoid HCTZ in postmenopausal women on estrogen. At least one study has shown that, among postmenopausal women, HCTZ was less well tolerated than moexipril and HCTZ is known to increase blood glucose while postmenopausal status by itself already increases insulin resistance and risk of developing diabetes mellitus. The differences in CVD effects of the various first-step drugs specifically among post-menopausal women on estrogen therapy have not been investigated and hence, will be investigated in this study.

Methods: The data of the ALLHAT was utilized to examine the effects of amlodipine and lisinopril compared to chlorthalidone in a subgroup of women on estrogen. These women were 55 years of age or older with hypertension and at least one other CVD risk factor from 623 North American centers (n=2752). ANOVA test and student t-test were used to compare the baseline characteristics between subjects in each group of therapy. Hazard ratios and 95% confidence intervals (CIs) were obtained from the Cox proportional hazards model to compare the clinical outcomes between the three antihypertensive groups. A composite endpoint of cardiovascular mortality and nonfatal myocardial infarction, stroke, coronary revascularization procedure, angina, heart failure, and peripheral vascular disease was used, as well as each CVD outcome individually and all-cause mortality.

Results: There were no significant differences in the baseline characteristics of the three treatment groups. Compared to chlorthalidone, the hazard ratios for the composite endpoint were 1.03 (95% CI 0.94-1.13, p-value=0.55) for amlodipine and 1.02 (95% CI 0.93-1.12, p-value= 0.64) for lisinopril. Likewise, hazard ratios for all-cause mortality as well as the individual CVD outcomes were not significantly different among either lisinopril or amlodipine compared to chlorthalidone.

Conclusion: Among women aged 55 years or above on estrogen, calcium channel blockers and angiotensin-converting enzyme inhibitors as initial antihypertensive therapy are associated with similar cardiovascular outcomes compared to thiazide diuretics. Therefore, for post-menopausal women on estrogen replacement therapy, physicians should not feel constrained to use HCTZ, but should instead choose initial anti-hypertensive medication taking into account individual patient’s side effect profile and other comorbidities.

References


Association of Chronic Pain and Myocardial Infarction

Authors: Jigar J. Patel MD, Talal Alzahrani MD, Shaneke Weerakoon MD, William Borden MD

Introduction: Chronic pain affects many patients and is the primary reason for visits to primary care clinics and emergency departments. In previous studies, chronic pain has been linked to mortality and cardiovascular disease. This study further explores this relationship by analyzing the association between chronic pain and myocardial infarction using a national database.

Methods: The National Health Interview Surveys of 2017 (n= 26,445) were used to examine the cross-sectional association between frequency of pain (never, some days, most days, every day) and myocardial infarction in a single logistic regression model that also included demographics [age, gender, Body Mass Index (BMI)] and health characteristics (smoking, hypertension, diabetes, hypercholesterolemia and exercise).

Results: Participants who reported having pain every day were more likely to be female (60% vs. 52%) and older (61 vs. 49 years) than participants who reported never having pain. The rates of daily smoking (23% vs. 9.6%), diabetes (24.9% vs. 8%), hypertension (63.1% vs. 29.2%), hypercholesterolemia (50.4% vs. 27.4%), and myocardial infarction (10.9% vs. 2.6%) were significantly higher in participants who reported having pain every day compared to subjects who reported never having pain. The logistic regression model showed that reporting daily experience of pain was independently associated with increased odds of having had a myocardial infarction (OR=2.19, 95% CI 1.69, 2.83, p <0.001) as was experience of pain most days (OR = 2.08, 95% CI 1.49-2.91, p <0.001), and experience of pain some days (OR = 1.51, 95% CI = 1.24-1.83, p <0.001) compared to never having pain after adjusting for demographics and health characteristics.

Conclusion: The frequency of chronic pain is significantly associated with the rate of myocardial infarction after adjusted for cardiovascular risk factors including age, gender, BMI, hypertension, diabetes, hypercholesterolemia, and lack of exercise. Future studies are needed to evaluate the efficacy of managing pain in reducing myocardial infarction.

References

DOMINICAN REPUBLIC RESEARCH POSTER FINALIST - PAMELA PINA SANTANA, MD

Cardio-Oncology Services in a Developing Nation: Patients Profiles and Practice Patterns

Authors: Pamela Piña, Amparo Taveras, Víctor A. Bueno, Cristal Vásquez, Gerardo De la Rosa, César J. Herrera.,
Institutional affiliations: CEDIMAT Cardiovascular Center, Santo Domingo, Dominican Republic

Introduction: The longevity of cancer patients is increasing owing to early detection but also due to improvements in antineoplastic therapies. As such, it is estimated that by 2026 there will be more than 20 million cancer survivors. Unfortunately, these advances have led to a 25% risk of cardiovascular mortality among this population since both entities share similar risk factors in addition to the fact that antineoplastic agents are fraught with significant cardiotoxic potential. As cardiovascular disease is a major cause of death among cancer survivors; developing countries are not exempt from the high prevalence of both pathologies. Limited data exists on local practices aimed at monitoring of cardiotoxicity in this population nor are there systematically applied protocols designed at prevention during and after antineoplastic therapy. We sought to determine cardio-oncological practice patterns and cardiovascular profiles of patients on antineoplastic therapy in the Dominican Republic through the creation of a registry.

Methods: A single institution registry of cancer patients over 18 years of age referred by their oncologist to the highest-volume echo lab in the country pre-chemo or on chemo were included for analysis of demographic, clinical, biomarkers and echocardiographic profiles. Investigators were not involved in their initial care; treating oncologists received no instructions. Referral and follow-up patterns where also measured. A Microsoft Office Excel program data sheet was created for analysis and recording. Data collection included: Left ventricle ejection fraction (EF), Global Longitudinal Strain (GLS), Brain Natriuretic Peptide and Troponin levels; presence of obesity (Body Mass Index ? 30), hypertension, diabetes, dyslipidemia, tobacco use, previously known coronary artery disease, type of cancer and anti-neoplastic treatment.

Results: From September 2016 to September 2018, 471 echocardiograms were performed in 309 patients: 72% were baseline studies and 28% (n=88) follow-up patients at mean 3-months. A total of 232 (75%) women, mean age 54 (18-85) years, 72% internally referred within our center. The most prevalent cancers were breast (59%), colon (7%) and lung (6%) treated with taxanes (22%) anthracyclines (19%) and trastuzumab (9%). Overall prevalence of cardiovascular risk factors was 71%, hypertension being the most common (68%). The mean baseline EF was 64% and GLS -19%. Cardiotoxicity was diagnosed in 11% (n=35) of follow-up patients with Δ baseline vs follow-up EF 59% (p = 0.0001) and Δ GLS -16% (p = 0.0162). Among patients with cardiotoxicity 72% (n=25) had at least one cardiovascular risk factor, being hypertension the most prevalent (51% n=18). Overall survival was 97%. Only 13% of the cohort (n=309) had cardiac biomarkers measured.

Conclusion: In this cohort, recently diagnosed cancer patients had a high prevalence of cardiovascular risk factors and a suboptimal pattern of monitorization. There is a need to create initiatives aimed at improving adherence to guidelines in developing nations.
Vinorelbine plus Capecitabine (Vinocap): A Retrospective Analysis in Heavily Pretreated HER2 negative Metastatic Breast Cancer Patients

Authors: Luis E. Aguirre; Alfredo Torres; Jeremy Ramdial; Ana Sandoval; Reshma Mahtani; Charles Vogel

Introduction: Even though the overall survival of primary breast cancer has improved significantly over the past 4 decades metastatic breast cancer (MBC) continues to be viewed as an essentially incurable entity associated with only 2.5% disease free survival at 15 years. Despite these numbers, palliative therapies have led to modest increases in 5-year survival and better quality of life. Notwithstanding the fact that hormonal treatment remains the mainstay of management for hormonally sensitive MBC, almost all patients will eventually need cytotoxic chemotherapy for palliation purposes. Specifically addressing cytotoxic therapy, single agent modalities have been preferred over combination regimens given the high degree of toxicity and restricted responses associated with their implementation. In heavily pretreated patients with increasingly limited options for palliative management the focus should be on ensuring proper quality of life.

With these facts in mind the authors conducted a retrospective analysis of a highly effective and minimally toxic combination regimen used in-house for over 2 decades, prompted by in vitro studies showing up-regulation of thymidine phosphorylase by vinorelbine to improve capecitabine effectiveness.

Methods: The investigators retrospectively analyzed a cohort of 67 women with human epidermal growth factor receptor (HER2) negative MBC treated at a large breast cancer community practice and a local cancer center with vinorelbine 22.5mg/m2 IV on day 1 and 8 combined with capecitabine 1 gram PO BID for 14 consecutive days of 21 day cycles. Patients were treated on average with 4 lines of chemotherapy. Data on clinical outcomes and patients characteristics were collected and evaluated.

Results: A total of 67 patients received the combination of vinorelbine with capecitabine and 2 patients among the 67 had two separate exposures giving an evaluable sample size of 69. Clinical benefit rate, defined as complete response, partial response or stable disease ≥ 6months, was 55.07%. 4.34% had a complete response, 18.8% had a partial response and 31.9% had stable disease for more than 6 months. Median progression free survival (PFS) time was 6.2 months and overall survival 35.47 months from start of VINOCAP therapy. The most common grade 3-4 toxicity was neutropenia in 10% of cases. Dose had to be reduced in 18% of the patients due to toxicity. The regimen was very well tolerated and side effects were rarely seen.

Conclusion: The combination of Vinorelbine with Capecitabine appears to be an active and well-tolerated regimen in women with MBC. A PFS of 6.2 months and clinical benefit in >50% of cases especially when used after ≥4 lines of chemotherapy with no reported instances of alopecia seem compelling arguments that should warrant consideration in this patient population. Consequently, Vinocap may serve as an additional lifeline in heavily pretreated patients confronting increasingly limited options for palliative management with preservation of quality of life.

References

Prognosis of Patients with Acute Kidney Injury after Percutaneous Mechanical Thrombectomy: a Retrospective Cohort Analysis

Authors: Hongchuan H. Coville1,2, Tien-Min Lee1,2, Hong Liang1, Paul Alfino1,3, Christopher Bray1,2, Matthew Calestino1,2,1University of Central Florida College of Medicine, Graduate Medical Education, Orlando, FL, 2North Florida Regional Medical Center, Internal Medicine, Gainesville, FL, 3 Nephrology Associates of North Central Florida, Gainesville, FL

Introduction: Percutaneous Mechanical Thrombectomy (PMT) is an endovascular procedure utilized in both arterial and venous thrombosis. Acute kidney injury (AKI) has been recognized as one undesired sequela of PMT. There is no study to investigate the outcomes among this specific patient population.

Methods: This is a retrospective cohort study using the de-identified clinically relevant datasets from inpatient database of seven community hospitals between 01/2015-12/2017. There were total 262 unique adult patients who underwent PMT included in the final cohort analysis. The primary outcome was in-hospital mortality which was the composite of recorded death during hospital stay or discharges to a hospice. The secondary outcome was the length of hospital stay (LOS).

Results: There are 28 out of 262 (10.9%) patients that developed post-PMT AKI. The composite in-hospital mortality was 21.4% among AKI group and 6.8% (P = 0.009) among non-AKI group. Among AKI group 19 (67.9%) patients stayed in the hospital longer than five days, and 115 patients (49.1%) among non-AKI group stayed longer than five days (P = 0.061). Univariate analysis showed that sex (P = 0.026), race (P = 0.044), shock (P = 0.028) and post-PMT AKI (P = 0.009) were correlated with the composite in-hospital mortality; and sex (P = 0.012), age (P=0.026), heart failure (P = 0.002), chronic kidney disease (P = 0.009), and diabetes (P = 0.031) were correlated with LOS. Among patients with LOS > 5 days 19 (14.2%) patients developed post-PMT AKI and 9 patients (7.0%) had post-PMT AKI among group with LOS ≤5days (P = 0.061). Using multivariate logistic regression analysis with Backward elimination method, two independent prognostic risk factors were revealed that significantly increased the risk of the composite in-hospital mortality: age (Odd Ratio [OR] = 1.07, 95% Confidence Index [CI] 1.03 - 1.12, P=0.001) and post-PMT AKI (OR = 4.77, 95% CI 1.42 – 16.11, P = 0.012). Using the same multivariate analysis model, among patients with post-PMT AKI, there was a trend of longer hospital stay though it is not statistically significant (OR= 1.86, 95% CI 0.77 – 4.46, P = 0.167).

Conclusion: Among patients treated with PMT in hospitals, the risk of dying is almost five times higher for patient who developed post-PMT AKI. Vigilant renal protection strategies are required before, during and post PMT procedure.

References

#InCtrl: the Impact of Sexual Education Programs on Today’s Youth

Authors: Edgar S. Fernandez MD, Angel Sanchez MD, Sasha Thiel MS3, Maraisha Philogene MS3, Michael DeDonno PhD, Allison Ferris MD

Introduction: In 2016, 39,782 individuals were diagnosed with HIV in the United States. According to the Center for Disease Control and Prevention, in 2016, adolescents between the ages of 13-24 represented up to 21% of all new HIV diagnosis in the country. In Broward County 15% of all individuals living with HIV were between 13-24 years of age. In November of 2017, Broward County was ranked number two in the U.S. of newly diagnosed HIV infections.

A lack of knowledge and increased risk-taking sexual behaviors’ amongst today's youth contributes to the spread of HIV. However, an increase in knowledge can alter sexual behavior within this population. Recent literature suggests that sexual education programs increase knowledge and positively influence lifestyle changes. Further insight into how specific HIV focused sexual education programs impact knowledge and risk-taking behavior may provide a template for future sexual education curriculums.

Methods: Over a one-year study, a sample of 102 individuals between 13-24 years of age in Broward County schools completed a one-hour HIV/AIDS educational session. The training session was created and delivered by representatives of the World Aids Museum and Educational Center, located in Wilton Manors. Prior to the educational session, participants completed an HIV knowledge pre-test. At the completion of the training session, participants were re-tested on HIV knowledge post-test.

Results: To explore the unique contribution of the training session, a paired-samples t-test was conducted. Specifically, the analysis determined whether there was a statistically significant mean difference between the pre-test and post-test. There were no outliers in the data as assessed by inspection of a boxplot for values greater than 1.5 box-lengths from the edge of the box. The assumption of normality was not violated, as assessed by Shapiro-Wilk’s test (p = .181). Participants performed better on the post-test (M = 17.39, SD = 2.11) than the pre-test (M = 15.07, SD = 2.40). The post-test elicited a statistically significant mean increase of 2.32, 95% CI [1.695, 2.946], t(102) = 7.359, p < .000. Further, as reported by Cohen (1988), results revealed a large effect, d = 0.725.

Conclusion: A significant improvement of knowledge about HIV occurred when participants were exposed to the sexual education program focused on HIV. Youths may benefit from HIV educational programs to further understand HIV, HIV stigma, and the implications of HIV infection. Future research could explore the influence of HIV focused sexual education on risky sexual behavior and the likelihood of contracting HIV.
Myocardial Infarction Risk Factors in Young Hispanics

Authors: Christopher Foth, DO, Josue Rizo, DO, Marc Iskandar, DO, Jose Paz, DO

Introduction: Very little research has evaluated myocardial infarction in young patients and even less data exists for young Hispanic patients. Classical risk factors do not appear to have the same magnitude of effect in Hispanic populations leading to the "Hispanic Paradox."

Methods: Data from Hispanic patients age 45 and under that presented with a myocardial infarction during a 5 year period at a community hospital in a predominantly Cuban-American area was analyzed. Charts were reviewed for risk factors and angiographic data. Data was analyzed for statistical significance using IBM SPSS.

Results: Dyslipidemia was the most common risk factor with a prevalence of 87.7% and low high-density lipoprotein was the most common abnormality (72.3%). Patients were also more likely to be male (81.6%), hypertensive (68.6%), obese (57.8%) and have a smoking history (50.7%). Men were more likely than women to present with an ST elevation myocardial infarction (52% vs 35%). Uninsured patients represented 37.6% of the population. These patients had similar coronary artery disease risk factors but were more likely than insured patients to abuse cocaine (13% vs 6%) and amphetamines (9% vs 3%), and to present with a total occlusion (73% vs 64%). Patients using amphetamines had an ST elevation myocardial infarction 88% of the time while cocaine use showed no statistically significant difference. Single vessel disease was present in 89% of patients with the left anterior descending artery being the most commonly involved vessel (48%). Patients presenting with multi-vessel disease (> 2 vessels) were more likely to have a higher body mass index, hypertension, and a family history of ischemic heart disease.

Conclusion: Dyslipidemia, hypertension, obesity and smoking are the most prevalent modifiable risk factors in young Hispanic patients presenting with a myocardial infarction. Early risk factor intervention should be encouraged in Hispanic communities to reduce the burden of ischemic heart disease.

References

A novel target for non-small cell lung cancer

Authors: Lingbin Meng, MD, PhD; Xuebo Yan, MD, PhD; Xiaochun Xu MD, PhD. Florida Hospital, Orlando, FL; MD Anderson Cancer Center, Houston, TX

Introduction: Raf kinase inhibitor protein (RKIP) is an inhibitor of Raf. By binding Raf, RKIP could inhibit Ras-Raf-MEK-ERK signaling, a major oncogenic pathway. Reduced RKIP expression is observed in many cancers including prostate cancer, breast cancer, etc. It is also recently identified RKIP may have anti-tumor and anti-metastasis properties. However, there is no published work about the role of RKIP in non-small cell lung cancer (NSCLC). In this study, we are the first to investigate RKIP expression in NSCLC patients and its correlation with tumor stage. Besides, our in-vivo experiments showed up-regulation of RKIP could be promising to treat NSCLC.

Methods: 32 NSCLC patients were included for this study with 6 at stage I, 8 at stage II, 7 at stage III and 11 at stage IV. Their cancer tissues were collected for analysis of RKIP expression and ERK signaling. Western blot and immunostaining were performed to detect expression level of RKIP protein, and phosphorylated ERK. Six-week-old C57BL/6 mice were purchased and maintained in the Laboratory Animal Center of our hospital. NSCLC cell line A549 was kindly provided by Dr. Xiaochun Xu, M.D. Anderson Cancer Center. This cell line was transfected with plasmid pIRES-Puro2-RKIP-1-Myc to up-regulate RKIP expression (experimental group) or with plasmid pIRES-Puro2-Myc for control group. Both groups contain 7 mice and each mouse receives subcutaneous injection of 5 x 10^5 cells on the left flanks. 4 weeks later, mice were sacrificed and tumor tissues were collected for tumor growth analysis. Also, similar experiments were performed with intravenous injection of 10^6 cells and the lungs were collected to study cancer metastasis.

Results: RKIP expression correlates with NSCLC tumor stages and expression of phosphorylated ERK. Late stages of NSCLC (III and IV) has a lower expression of RKIP than early stages (I and II) (P<0.01). Lower expression of RKIP is also accompanied with higher expression of phosphorylated ERK. The in-vivo experiments showed up-regulation of RKIP could inhibit tumor growth and metastasis.

Conclusion: This is the first study to demonstrate RKIP plays an oncolytic role in NSCLC. Our results revealed low expression of RKIP correlates with late stages of NSCLC and high level of p-ERK. So, RKIP is a good prognosis indicator for NSCLC. Besides, up-regulation of RKIP can inhibit NSCLC growth and metastasis, and therefore RKIP is a promising novel target for NSCLC. Our future study is focused on designing a virus carrying RKIP for treatment of NSCLC.
The effect of early nutritional intervention on nasopharyngeal carcinoma treated with chemoradiotherapy

Authors: Lingbin Meng, MD, PhD; Xin Jiang, MD, PhD; Robin Krimm, PhD.

Introduction: Nasopharyngeal carcinoma (NPC) is the most common cancer originating in the nasopharynx. Patients with NPC frequently had the problem of malnutrition at the time of diagnosis. Chemoradiotherapy (CRT) can even worsen the situation. Therefore, nutritional intervention should be applied to prevent CRT-associated weight loss and interruption of CRT. However, it is still controversial if early nutritional intervention is more beneficial than the late. This study is to investigate the influence of early nutritional intervention on patients with NPC by observing change of body weight, radiation-induced oral mucositis and treatment tolerance.

Methods: From March 2017 to January 2018, 54 patients with NPC were randomly assigned into early nutritional intervention group (28 cases) and late group (26 cases). Both groups received CRT for 7 weeks and patients’ data was collected on the first day of CRT, the last day of CRT, and 3 months after ending CRT. The early group received enteral nutrition on the first day of CRT while the late group wouldn’t receive enteral nutrition until they developed problems with oral dietary intake. The collected data included age, sex, tumor stage, weight change, incidence of advanced mucositis, number of patients with CRT breaks, and days of CRT delayed for toxicity. The data were expressed as average±SD. The relation between the single variables was analyzed using the chi-squared test for categorical variables. Differences in continuous variables between groups were compared using the Student t test for unpaired data. P<0.05 was accepted as statistically significant.

Results: Both groups lost weight at the end of CRT and 3 months after the end of CRT. However, at the later time point, the early group started to regain their weight while the late group continued to lose weight. At both time points, the early group had less weight loss than the late group. Also, the early group showed lower incidence of advanced mucositis (III,IV ) and higher treatment tolerance than the late group.

Conclusion: Early nutritional intervention provides beneficial outcomes by reducing weight loss, decreasing advanced mucositis and increasing CRT treatment tolerance. This finding demonstrates early nutritional intervention is important to maintain patient’s nutritional status and has crucial clinical significance. This study stands out because it is randomized, prospective and double-blinded.
Survival after Liver transplantation for Alcoholic Cirrhosis with and without Hepatitis C

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Introduction: Liver transplantation (LT) is a life-saving intervention for end-stage liver disease due to alcoholic cirrhosis (AC). Several published studies have demonstrated favorable survival outcomes following LT for AC. However, data related to outcomes following LT for AC among patients who have coexistent chronic hepatitis C infection is limited.

Methods: We retrospectively reviewed 2091 recipients of deceased-donor LT at an academic transplant center from January 2000 to December 2012. We included recipients of primary whole liver alone transplants and those with coexistent chronic hepatitis C infection and alcoholic cirrhosis. Recipients of prior transplants, multi-organ transplants and cholangiocarcinoma cases were excluded. Kaplan-Meier survival analysis was performed comparing LT recipients in two groups: AC with and without chronic hepatitis C infection.

Results: 456 LT recipients met the inclusion criteria. Mean age 60±7 years; mean BMI at listing 29±6; 61% male and 20% had HCC. Mean MELD score was 19±8; mean donor age was 29±6. There was no statistical difference between LT recipients in the 2 groups in terms of donor age, donor BMI, MELD score, gender, and cold or warm ischemia times (p>0.05). Median overall survival was comparable between two groups (4.1 years vs 3.9 years, log-rank p=0.2).

Conclusion: Liver transplantation in patients who have alcoholic cirrhosis and chronic hepatitis C infection has comparable survival outcomes to those who underwent LT for AC alone. Patients with alcoholic cirrhosis benefits well from liver transplantation regardless of the hepatitis C status.
High-risk opioids use pattern of chronic non-cancer pain patients in a resident continuity clinic

Authors: Luwei Tao MD, Mohammed Wazir MD, Maham Jalil MD, Akriti Jain MD, Mamoon Ur Rashid MD, Manoucher Manoucheri MD

Introduction: Overuse and misuse of opioids is a national health crisis. More than 72,000 people died from drug overdose in the US in 2017 and two-thirds of the death was attributed to opioids use. The epidemic of drug overdose is fueled by high-risk pattern of opioids prescription for chronic pain. In order to improve the safety of pain management and reduce long-term opioids therapy associated complications, CDC implemented the new Guideline for Prescribing Opioids for Chronic Pain in 2016. In compliance with this guideline, our clinic implemented a new opioids prescription policy. The goals of our project are (1) to characterize high-risk prescription pattern for non-cancer chronic pain patients who get opioids prescriptions from our clinic and (2) to compare the change of prescription practice pattern after the implementation of our new opioids policy.

Methods: The study was initiated by chart review of patients seen in a resident continuity clinic from 6/1/2016 to 2/27/2017. Inclusion criteria for chronic opioids-dependent patients included: opioid use consecutively for more than 3 months; daily dose more than 0.8 tablet of any opioid; and pain management for non-cancer conditions. The daily dose morphine milligram equivalent (MME), types of opioids and co-prescription of benzodiazepine were determined. High risk prescription pattern was identified as high MME (higher than CDC recommendation of 50 MME), co-prescription of two or more opioids, co-prescription of benzodiazepine. Our ongoing interventions included; new agreement for controlled prescription; naloxone with frequent reassessment if daily dose >50MME; and pain management referral if daily dose>90MME. Same group of patients will be reevaluated for opioids consumption 6 months after implementation of new opioids policy.

Results: A total of 92 patients were identified as chronic opioids-dependent patients. Hydrocodone and Oxycodone were the two most common prescribed opioids in our clinic, followed by Tramadol. Among this group of 92 patients, 66 of them had daily opioids dose less than 50MME; 7 had daily opioids dose between 50-90MME and 19 had daily dose more than 90 MME. Mean daily dose was 55.8MME. Of the same group patients, 30.4% took more than 2 types of opioids and 20.7 % consumed both opioids and benzodiazepine.

Conclusion: A significant portion of patients on chronic opioids in our clinic have high risk features, including high daily dose more than 50 MME, taking multiple opioids or taking both opioids and benzodiazepine. Intervention is necessary to improve the safety and effectiveness of pain management and reduce the risks of serious complications associated with chronic opioids therapy.

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Retrospective Analysis of the Use and Yield of Radiologic Studies in IPMN Surveillance

Authors: Cicily Vachaparambil, MD, Neil Kapil, MD, Jennifer Xu, MD, Chunsu Jiang, MD, Steven Keilin, MD, Field Willingham, MD, Qiang Cai, MD

Introduction: Intraductal papillary mucinous neoplasms (IPMNs) of the pancreas are diagnosed with increasing frequency as incidental findings on abdominal MRI or CT, often on imaging studies done in the hospital for other indications. Close surveillance with imaging is indicated according to most recent guidelines, which are based on weak evidence. This study evaluates utility of radiologic studies in the natural history of incidental pancreatic cysts and cost-benefit of close surveillance.

Methods: Of 1167 patients with a diagnosis of IPMN between 10/2010-02/2018 at our institution, 150 were randomly selected for retrospective chart review. Exclusion criteria included known pancreatic cancer or other reason for routine imaging. Charts were reviewed for outcomes including age discovered, whether incidental or not, lesion features, amount of growth, time followed, biopsy results, number and type of imaging studies done, and eventual outcome.

Results: Out of 141 patients that qualified, most were female (57%) and IPMN was discovered at 60-70 years (32.6%). Of all IPMNs, 69.5% were incidentally found with 73.8% on MRI. Initial IPMNs were most likely 6-10 mm and side branch lesions. 83.7% of these did not have suspicious features, defined as dilation of the main pancreatic duct >10 mm, size >30 mm, or a solid component. 66.7% were asymptomatic, without abdominal pain, nausea, or jaundice prior to imaging. Despite this, lesions were followed over an extended time course, with 14.1% longer than 5 years. Most of these IPMNs, 65.2%, did not have any changes and remained stable, 24.8% had some growth at an average of 1 cm, 9.2% were benign on biopsy, and 2.8% were confirmed to be cancer. In total, 360 MRIs and 97 CTs were done for these 141 patients to follow IPMNs. For non-suspicious lesions, an average of 2.96 follow-up studies were done per person. This equates to 60 CTs/person and 2.38 MRIs/person. For lesions with suspicious aspects, an average of 4.52 studies were done per person. This equates to 1.13 CTs/person and 3.43 MRIs/person.

Conclusion: Our study supports more conservative use of imaging in IPMN surveillance. As many of these lesions are found on radiologic studies done for other indications in the hospital, careful decision making about further surveillance are warranted given the extended months of follow-up, number of imaging studies, and a majority of outcomes showing no change or minimal growth. With more conservative use of imaging, costly follow-up procedures, including ERCP, biopsy, and surgery, and adverse effects of radiation can be avoided. We recommend further studies across training sites to improve surveillance guidelines to be both cost effective and maintain quality patient care.

References

A Key to Lock the Revolving Door: A Residency Program Initiative to Transform Transitions of Care

Authors: Catherine Apaloo MD. Jacob Barry MPH

Introduction: Readmissions to hospitals cost the United States over $17 billion annually, with $12 billion of this amount being spent on potentially avoidable readmissions. As information is not always transmitted to the primary care provider, which in turns results in sub-optimal care. Transitions of care is an ACGME Milestone, however, it is not necessarily a part of the residency curriculum. Numerous interventions have been proposed to address this issue. In our program we designed and implemented a resident led transition of care team.

Methods: We implemented a multi-component integrated team-based, transition-of-care service intervention and assessed its impact on show rates for TOC visits and 30-days readmission rate.

The 3 components of the intervention starts at the time of discharge by a call to the TOC hotline. The second process is utilizing a call by the TOC lead resident within two business days of discharge which includes medication reconciliation, referrals and social needs. The final step is a face to face TOC visit within 7 to 14 days of discharge with a resident, a pharmacist and a social worker.

We conducted a retrospective chart review of Piedmont Athens Regional Medical Center patients discharged and referred to the TOC residency clinic by calling the TOC hotline prior to discharge. We measured the 30-day readmission rate, and TOC visit show rate. 150 patients were included from December 19, 2017 to March 1, 2018. Data analysis was done using SPSS.

Results: The 30-day readmission rate was 9% for the 150 patients, among the 66 patients who received all components of the intervention the readmission rate decreased to 3%.

The show rate among patients who received the follow up phone call was 90% in comparison to 28% for patients whom we were unable to reach. The general show rate for our clinic for 2017 was 73 %.

The process is ongoing, with 3 months data analysis. Future plan includes TOC home visits and mobile clinic to address areas of high utilization of health care.

Conclusion and Implications for Patient Care: The initiative led to building a TOC experience for internal medicine residents. It established the process, provided easy access though the hotline to ensure proper transitioning from the inpatient to the outpatient setting. And more importantly it increased our TOC visit show rate. The calculated cost saving was estimated to be 780 000 $ in 3 months period. This method could be implemented in other residency programs to emphasize the culture of safe transitions.
Shared decision making in diverse patient populations: relations to health literacy and health autonomy

Authors: Tanya Reddy, M.D; Neha Naithani, M.D; Jessica J. Shotwell, M.S; Zahraa Rabeeah, M.D; Laurel Murrow, M.D; Lisa M. Renzi-Hammond, Ph.D

Introduction: Shared decision making (SDM) is the gold standard for patient communication in many healthcare settings. Although, research suggests SDM is rarely practiced. There are many possible reasons for inconsistent application of SDM, including poor health literacy and low autonomy on the part of the patient, and lack of hands on training in autonomy support for healthcare providers. The purpose of this quality improvement initiative is 1) to characterize the health literacy and autonomy levels of patients in a large resident training clinic affiliated with a major hospital system and medical school; 2) to determine the extent to which the medical residents responsible for patient care believe they are successfully engaging in SDM; 3) to determine the extent to which patients and residents align on their perceptions of SDM engagement; and 4) to identify targets for intervention to improve SDM in this environment.

Methods: Health literacy and health autonomy were measured using a validated questionnaire given either verbally (n = 70) or in writing (ongoing, n = 26), while patients waited for their appointments. Following the patient appointment with the medical resident, patients and residents were independently measured regarding their perceptions of SDM during the visit. Neither residents nor patients were privy to the post-visit perceptions of the other party.

Results: Patients varied widely in age (M= 47.0 ± 13.9 years) and racial and ethnic diversity (n = 44.3 % Caucasian, 45.7% African American). Health literacy varied widely between patients. While patients felt confident in their ability to navigate the clinic environment and read signage and medical forms, they also tended to report needing significant help in answering questions and some difficulty in reading medication labels. With respect to health autonomy, patients felt a strong desire for autonomy supportive behaviors from their physicians, including asking for input and addressing lifestyle concerns as part of the treatment regimen. Patients and physicians varied in their perceptions of the extent to which SDM was used to support patient autonomy.

Conclusion: SDM is the gold standard for patient communication in most medical settings, but it is inconsistently practiced. Understanding the extent to which patient health literacy and feelings of autonomy relate to successful application of SDM will allow physicians to intervene to improve communication, particularly in patients with low health literacy.
HAWAII RESEARCH POSTER FINALIST - YOUNG SOO RHO, MD

Associated Factors and Survival Implications of Biopsy Diagnosis of Hepatocellular Carcinoma.

Authors: Young Soo Rho M.D., Jared Acoba M.D. University of Hawaii Internal Medicine Residency Program.

Introduction: Hepatocellular carcinoma (HCC) is one of the few cancers that can be diagnosed based on image findings alone. However, biopsy of HCC can confirm histologic subtype, and potentially be used to identify novel therapies. The factors associated with undergoing a biopsy diagnosis and the difference in survival between patients diagnosed with imaging versus biopsy have never been studied.

Methods: We collected demographic, diagnostic, treatment, and survival data of 171,013 patients diagnosed with HCC between 2004 – 2015 from the National Cancer Database. To determine factors associated with undergoing a biopsy, binary logistic regression was performed. Univariate and multivariate cox proportional hazards regression models were created to determine impact of diagnostic method on survival. Variables included were race, sex, age, comorbidity, facility type (academic vs community), insurance, tumor size, presence of metastatic disease, alpha fetoprotein, total bilirubin, and administration of therapy. Analysis was performed with SPSS v25.

Results: We included 160,517 patients in the final analysis. The median age was 62 (18 – 90), 73.9% were male and 74.1% were white. 11.5% of tumors were 2cm or smaller and 13.7% of HCC were metastatic. 78,485 (47.7%) underwent a biopsy. In a multivariate model, black patients (OR 1.093; 95% CI 1.061 – 1.126), older patients(OR 1.579; 95% CI 1.537 – 1.622), larger tumor size (OR 3.208; 95% CI 3.085 – 3.335), private (OR 1.129; 95% CI 1.094-1.164) or Medicare insurance(OR 1.056; 95% CI 1.021 – 1.093), and metastasis (OR 1.318; 95% CI 1.275 – 1.361) were associated with biopsy diagnosis. Factors associated with an imaging diagnosis were female (OR 0.964; 95% CI 0.941 – 0.987), Asian/Pacific Islanders (OR 0.686; 95% CI 0.657 – 0.717), higher comorbidity index (OR 0.578; 95% CI 0.559 – 0.597), treatment at an academic center (OR 0.447; 95% CI 0.436 – 0.458), hyperbilirubinemia (OR 0.807; 95% CI 0.784 – 0.831) and elevated AFP (OR 0.571; 95% CI 0.549 - 0.594). Over the examined study period 2004-2015, imaging diagnosis was increasingly used. Patients who underwent biopsy demonstrated inferior survival with a HR 1.400 (95% CI: 1.385 – 1.417). After adjusting for other prognostic factors in a multivariate cox analysis model, biopsy had a much smaller impact on survival HR 1.017 (95% CI: 1.004 – 1.030).

Conclusion: Although imaging alone can be adequate to diagnose many cases of HCC, nearly half of the cohort underwent a biopsy. While the diagnosis by imaging was more frequently used in recent years, there were still racial and institutional differences in pattern of care. Establishing a diagnosis with a biopsy did not have a significant impact on survival.
Waking Up to New Guidelines: ASV Therapy for Central Sleep Apnea in Patients With Heart Failure

Authors: Dr. Sumir Brahmbhatt (Associate), Dr. Kevin Brown (Associate), Dr. Rajesh Daniels, Dr. Jordanna Hostler, FACP

Introduction: Central sleep apnea syndrome (CSAS) with Cheyne-Stokes breathing is an independent risk marker for poor prognosis and death in patients with heart failure (HF). Adaptive servo-ventilation (ASV), a noninvasive ventilatory therapy that delivers servo-controlled inspiratory pressure support on top of expiratory positive airway pressure, effectively alleviates CSAS. Evidence from a large randomized trial (SERVE-HF), however, demonstrated an increase in cardiovascular and all-cause mortality in HF patients with a reduced ejection fraction (EF) receiving ASV therapy compared to a control group. This led to a 2016 recommendation by the American Academy of Sleep Medicine (AASM) contraindicating ASV in HF patients with EF <45% and moderate-severe CSAS. In light of this recommendation, there was a need to determine if any of our clinic’s ASV patients have an EF <45% and intervene, as well as to establish a protocol to prevent patients with HF with EF <45% from starting ASV therapy.

Methods: A list of patients who had been prescribed ASV therapy for CSAS at Tripler Army Medical Center Sleep Clinic was compiled. A thorough chart review of each patient was performed to identify those who had (1) sleep-disordered breathing with >50% central apneic events, (2) chronic HF >12 weeks, (3) NYHA III/IV symptoms or NYHA Class II with 1 or more hospitalizations in past 2 years, and (4) EF <45% as determined by transthoracic echocardiogram, radionuclide ventriculography, or cardiac MRI. Those identified were then scheduled for an appointment to discuss the risks of and alternative treatments for ASV. Additionally, a standard operating procedure (SOP) is in the process of development to screen all new ASV candidates for heart failure and obtain cardiac evaluation for suspected HF or known HF without recent follow up.

Results: 117 patients were found to be currently prescribed ASV therapy; of these, 10 patients were found to have met the inclusion criteria. These individuals were contacted by phone and are currently being titrated off of ASV therapy. In December 2018, a 12-month progress analysis will be completed to follow up on these patients. A finalized SOP for HF screening for these patients was made and all physicians were notified.

Conclusion: This project effectively identified 8.5% of all patients on ASV therapy in whom ASV is contraindicated and dangerous. These patients were prescribed ASV before the results of the SERVE-HF trial came out and the updated AASM recommendations were made. By titrating them off of ASV and creating an SOP that screens and surveils ASV candidates for HF with reduced EF, we may avoid unnecessary increase in cardiovascular and all-cause mortality.
Useful or Futile? The Safety and Efficacy of Fecal Microbiota Transplantation in Elderly Individuals

Authors: Faisal S. Ali, MD, Zimu Gong, MD, Muhammad Akrmah, MD, Hamzah Abu-Sbeih, MD

Introduction: Worldwide, 70-80% of *Clostridium difficile infections* (CDI) are documented in elderly individuals. Besides being of older age, elderly patients suffer from multiple comorbidities, have impaired immunity, and are more likely to be residing in long term care facilities; all risk factors for a recurrent CDI (rCDI)\(^1\). Fecal microbiota transplantation (FMT) has been approved to treat severe, refractory and recurrent *clostridium difficile* infection (CDI), with additional investigations underway to expand the indications of FMT. However, data regarding the safety and efficacy of FMT in adults aged ≥ 65 is limited. A systematic review and meta-analysis of proportions was conducted to evaluate clinical utility of FMT in the elderly population.

Methods: A comprehensive search of Medline, Embase, Scopus, and Cochrane Library for studies from January 2010 to May 2018 was performed using controlled vocabulary as well as natural language terms for FMT and gastrointestinal disease. Studies enrolling patients aged ≥ 65 and reported the clinical outcome of CDI in elderly patients following FMT were included. Pooled rates [with 95% confidence interval (CI)] of clinical success, CDI recurrence after initial FMT, and adverse events (AE) associated with FMT were calculated. The quality of included studies was assessed. A logistic-normal random-effects model was applied to account for effect sizes close or equal to one. Inter-study heterogeneity was assessed.

Results: Overall, 749 initially screened studies yielded 9 studies (seven retrospective, two prospective) enrolling 361 elderly patients, of which, 212 (59%) were female. All patients had failed at least one course of antibiotic therapy prior to FMT. Overall, 309 patients responded to a single FMT. The pooled response rate after a single FMT was 90% (95% CI, 77-96%; \(\chi^2 = 11.64; d.f. = 7, P = < 0.001; \text{Tau}^2 = 1.24\), with a pooled rCDI rate of 7% (95% CI, 2-22%; \(\chi^2 = 19.54; d.f. = 7, P = < 0.001; \text{Tau}^2 = 2.22\) post FMT. Concerning the safety of FMT, 23 AEs were reported with a pooled AE rate of 5% (95% CI, 2-16%; \(\chi^2 = 4.60; d.f. = 7, P = 0.016; \text{Tau}^2 = 1.67\)). Two mortalities occurred due to intra-procedural aspiration with a gastroscopic FMT approach. Both the patients were older than 85 years of age and suffered from multiple comorbidities. All other AEs were mild, transient in nature, and did not require intervention. The mean follow-up duration was 16 months.

Conclusion: A single FMT is safe and efficacious for eradicating CDI in elderly patients, though some patients may require more than one FMTs. Caution should be practiced with a gastroscopic approach in patients with multiple comorbidities and those considered to be at high risk of aspiration. Future large scale studies that account for confounding factors are required to validate these findings.

References

Reducing Inappropriate Telemetry Utilization on an Internal Medicine Teaching Service

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Introduction: Inappropriate cardiac telemetry creates increased expense and higher workloads for medical staff. Hospitals are also limited on the availability of cardiac telemetry, which as a result can restrict patient flow. Revised clinical practice recommendations on appropriate telemetry use were released in 2017. We conducted a project aiming to reduce inappropriate telemetry use and improve patient flow within the hospital.

Methods: The study was conducted at a 300-bed urban teaching hospital. Baseline data on appropriate telemetry use was collected by manual review of non-intensive care patients on the Internal Medicine (IM) services for two weeks. Appropriate telemetry use was evaluated on the initial indication for telemetry and subsequent duration of telemetry use. The intervention was two educational case-based conferences given to all available IM residents and one informational session given to IM core faculty members. Post-intervention, there was a 2-week transition period prior to restarting data collection for 1 month. A cost analysis was performed based on differences between telemetry and standard rooms of an estimated $300/day. For the secondary outcome of patient flow, transfers out of the Intensive Care Unit (ICU) were evaluated for potential delays. ICU transfer delays were identified by manual chart review. Delays due to bed availability were classified as telemetry or general medicine floor (GMF) and quantified in 12 hours shifts. Transfer delays were evaluated for 2 weeks prior to the intervention and 6 weeks non-continuously following the initial intervention.

Results: A total of 960 patient days, 383 pre-intervention, and 577 post-intervention, were evaluated for telemetry appropriateness. The average proportion of patients inappropriately on telemetry significantly decreased from pre-intervention to post-intervention (67% to 57%, p=0.002), which corresponded with a decrease of 3.3 patients/day with inappropriate telemetry indications (See Figure 1). The overall number of patients on telemetry per day significantly decreased from 25.5 to 21.4 (p=0.006). The estimated cost savings from the intervention was $1230/day or $448,950 annually. For the secondary outcome of patient flow, the proportion of hours of delay due to telemetry bed availability compared to GMF significantly decreased (64% vs. 41%, p <0.001) following the intervention. The average transfer time delay due to telemetry bed availability per day trended downward (7.2 vs. 4.7, p=0.36). The overall transfer time delay out of the ICU did not change from before to after the intervention (11.2 vs. 11.5, p=0.95).

Conclusion: A brief educational intervention significantly decreased inappropriate telemetry use for patients cared for by IM residents. Time delays in transfers from the ICU to telemetry beds improved following resident education but did not change overall time in transfer delays. Further study is needed to evaluate the sustainability of the intervention and hospital-wide effects of implemented practice changes on patient flow.
Open Access Scheduling for Routine and Urgent Appointments to Cardiology Clinic

Authors: Abdisamad M. Ibrahim MD, Mohammad Al-Akchar MD, Bishal Bhandari DO, Hadi Mahmaljy MD, Basma Al-Bast MD, Mirza Ali MD, Cameron Koester DO, Abhishek Kulkarni MD

Introduction: In the past, when a primary care provider wanted to refer a patient to the SIU cardiology clinic, the referring provider or the nurse will have to call the central call center and request an appointment for the patient. However, this process was cumbersome and resulted in delays, jeopardizing patient safety and negatively impacting patient/ provider satisfaction.

Methods: SIU Healthcare created a Patient Access Center (PAC) in January 2017 to improve patient access to the SIU Cardiology (and other specialties). Guidelines were developed with the involvement of the key stakeholders. SIU Cardiology specified times on the provider schedules for PAC use only, noted all providers could see all cardiology diagnosis, and also identified sub-specialty interests for each provider. As per protocol, when a patient needs to see a cardiologist the primary care providers will put in a referral task in the EMR. The PAC team will then schedule that appointment according to the time slots provided to them by the cardiology clinic. To maintain/ sustain this program, The PAC team meets with SIU Cardiology team every four weeks to monitor the progress of the referral system.

Results: Our main outcome measure was the time between electronic referral to PAC to the time when an appointment was scheduled. As the project went on, the time difference was noticed fairly quickly and was downtrending. At the beginning of the project (January 2017), the average time for a routine referral to appointment set was 40 days. At the end of the project (April 2018), the average time from referral to appointment was down to 14.8 days. For urgent referral, average time to appointment was 49.3 days in the beginning (February 2017), however, at the end of the project (April 2018), it was down to 4.7 days.

Conclusion: The SIU Healthcare identified the problem of scheduling delays in important appointments in specialties, such as Cardiology. The PAC team was created to address this problem. With guidelines and input from SIU Cardiology, the PAC team was able to reduce the number of scheduling delays significantly.
Evaluation of the Neutrophil-to-Lymphocyte Ratio role in the prognosis of Severe Alcoholic Hepatitis

Authors: Yazan Abu Omar, Rohit Agrawal, Tejinder Randhawa, Yuchen Wang, Sanjay Patel

Introduction: Prednisolone is the cornerstone treatment for severe alcoholic hepatitis, but it increases the risk of infection. Multiple prognostic models have been proposed to early stratify patients into responders versus non-responders; the best so far is Lille score at day 4. We evaluated the use of neutrophil lymphocyte ratio in conjunction with day-4 Lille score.¹,²

Methods: We retrospectively reviewed the electronic medical records of patients who were diagnosed with severe alcoholic hepatitis with a Maddrey’s discriminant factor (DF) >32. Receiver-operating characteristic (ROC) curves were plotted for Day 4 NLR and Day 4 Lille score for prediction of 90-day mortality, and optimal cut-off values were determined. Patients were then subcategorized into groups based on the optimal cut-off values, and mortality rate in each group compared with Chi-square test to further validate the categorization. Finally, we performed multivariate analysis for prediction of 90-day mortality using Day 4 Lille score and Day 4 NLR, and constructed new prediction score based on odds ratio. The ROC curve of the new score was plotted and area under curve (AUC) was reported in comparison with pre-existing validated scores.

Results: We identified 104 patients with a confirmed diagnosis of severe AH requiring prednisolone therapy. Maddrey’s Discriminant Function for the included patients averaged at 65.2 with a standard deviation (SD) of 31.9. Upon Day 4 of hospitalization, Lille score averaged at 0.33 with a SD of 0.29; and NLR averaged at 10.5 with a SD of 10.7. In univariate logistic regression, both Day 4 NLR and Lille score significantly predicted 90-day mortality (p=0.049, p<0.001, respectively). We constructed the Day 4 Lille-NLR using the reported odds ratio in multivariate analysis as weight, and again plotted the ROC curve. The AUC for Day 4 Lille-NLR score is 0.889, higher than that of Lille score and NLR independently.

Conclusion: NLR is an easy/bedside test that you can calculate from CBC alone and has comparable (albeit less) prediction of mortality, outcomes assessment. Adding NLR to Lille (“modified” Lille score) adds increased performance characteristic in prediction of outcomes/mortality to the pre-existing model.

References


ILLINOIS RESEARCH POSTER FINALIST - BISHAL BHANDARI, DO

General Anesthesia versus Conscious Sedation in Transcatheter Aortic Valve Replacement

Authors: Bishal Bhandari DO, Alexander Worix BS, Manjari Regmi MD, Abdisamad Ibrahim MD, Cameron Koester DO, Priyanka Parajuli MD, Basma Al-bast MD, Mohammad Al-Akchar MD, Michael Buhnerkempe PhD, Abhishek Kulkarni MD

Introduction: Transcatheter Aortic Valve Replacement (TAVR) is an established alternative to surgical aortic valve replacement (SAVR) when treating symptomatic aortic stenosis in intermediate and high risk patients[1]. Historically, general anesthesia (GA) has been the primary form of patient sedation during TAVR. Recently, conscious sedation (CS) has been emerged as a safe option for TAVR. Under conscious sedation, respiratory drive remains intact, patients are safely able to maintain their airway, and brainstem reflexes continue to be functional. To date, there is no randomized controlled trial that contrasts the outcomes of general anesthesia vs. conscious sedation in this setting. However, there have been a few previous studies that reported no differences in mortality yet reduced hospital stays for patients [2,3]. While there is little data available comparing these two modalities, it is appropriate to explore variables that reflect the outcomes of patients who used either approach.

Methods: To determine the potential benefits of using conscious sedation compared to general anesthesia, 3 variables were compared in patients experiencing each method: (1) Total hospital length of stay (LOS), (2) ICU LOS, and (3) occurrence of adverse events (AEs) during hospitalization. To control for potential confounding factors, we included age, sex, atrial fibrillation, history of CABG, and PVD. All analyses were carried out in R statistical software.

Results: Hospital LOS was found to have a correlations with anesthesia method. CS was correlated with 69% shorter hospital stays compared to GA. ICU LOS was found to have correlations with history of CABG, PVD and anesthesia method. Patients receiving conscious sedation had 54% shorter ICU stays. Those with PVD had 101% longer ICU stays. Those with a history of CABG had 87% shorter ICU stays. No variables, including anesthesia method, were significantly associated with occurrence of AEs, indicating that the risk of complications was similar for both anesthesia methods.

Conclusion: Once addressing the outcome differences in terms of general anesthesia vs conscious sedation, several correlations were discovered. Conscious sedation showed shorter hospital LOS and ICU LOS. Patients with PVD had longer ICU stays. Anesthesia method showed no relationship with occurrence of adverse effects. Overall, conscious sedation compared to general anesthesia could potentially be the better alternative for TAVR.

References

Imaging Wisely: An Introduction to the ACR Appropriateness Criteria and Analysis of its Impact on Internal Medicine Residents.

Authors: Mike KW Cheng, MD; Andrea L Magee, MD; Carina W Yang, MD; Joyce W Tang, MD, MPH

Introduction: Inappropriate radiological exam ordering contributes significantly to healthcare waste. The American College of Radiology’s (ACR) Appropriateness Criteria (AC) were designed to inform radiological exam ordering practices, but many internists are unfamiliar with them. To promote high-value ordering practices and use of this evidence-based resource, our team of internists and radiologists developed a Choosing Wisely® inspired curriculum introducing internal medicine residents to the ACR AC.

Methods: We piloted our curriculum with University of Chicago internal medicine interns from 2017-2018. The curriculum included: (1) a discussion about medical risks of overuse of radiological exams and sample exam charges; (2) an introduction to the ACR AC website and mobile app; (3) application of the AC to case vignettes through Radiology-TEACHES™ software; (4) group discussions about the vignettes; and (5) a radiologist-led session about principles of high-value radiological exam ordering. Radiology-TEACHES™ tests learners with case vignettes and provides feedback on image ordering decisions with snapshots of relevant AC tables.

We used pre- and post-intervention surveys to assess for change in knowledge (through open-book testing using Radiology-TEACHES™ vignettes) as well as change in attitude related to high-value exam ordering. A follow-up survey given 3-11 months post-intervention assessed for durability of change. We utilized McNemar’s tests and paired t-tests for analysis.

Results: To date, 23 interns completed the curriculum and pre- and post-intervention surveys; 18 completed the follow-up survey. Pre-intervention, 35% of residents were unaware of the ACR AC and only 25% reported it among their top 3 resources guiding ordering decisions. Post-intervention, knowledge scores for appropriate exam ordering increased from 59% correct answers (SD 0.16) to 89% (SD 0.14); p=0.0001. The AC were more frequently referenced during post-intervention assessment (17% of residents pre vs. 74% post; p=0.0003). Post-intervention, more residents felt comfortable with their knowledge of exam costs (4% pre vs. 74% post) and discussing cost with patients (9% pre vs. 61% post), and more residents valued the ACR AC (65% vs. 96%); p-values all <0.05. All residents found the curriculum helpful; 91% planned to change their ordering practices. At 3-11 months post-intervention, 78% referenced the ACR AC at least monthly and 67% reported changing ordering practices because of the curriculum. Knowledge scores remained significantly improved.

Conclusion: This curriculum was well-received and produced durable change in knowledge and attitudes towards high-value radiological exam ordering and use of the ACR AC. It is low cost and can be easily disseminated. It is timely as the Protecting Access to Medicare Act will require clinicians to reference appropriate use criteria (AUC) to justify ordering advanced imaging studies and the AC are approved by the Centers for Medicare and Medicaid to provide AUC. Lastly, given the reported sustained change in ordering practices as a result of the curriculum, it has strong potential to influence downstream ordering practices.

References

Trends related to sudden cardiac arrest

Authors: Dipesh Ludhwani M.D., Elizabeth Retzer M.D., Karam Khaddour M.D., Noemi Arias Ph.D.

Introduction: Sudden Cardiac Arrest (SCA) is defined as a spontaneous cessation of cardiac activity leading to unresponsiveness and loss of circulation. Based on 2016 Resuscitation Outcomes Consortium (ROC)-Cardiac Epistry statistics, survival rates for Out-of-Hospital Cardiac Arrest (OHCA) and In-Hospital Cardiac Arrest (IHCA) were 12% and 24.8% respectively. SCD outcome may depend on a variety of factors ranging from individualized patient risk status to quality of resuscitation. This paper aims to study the role of such elements and to establish trends among patients with SCA.

Methods: A retrospective chart review of 81 patients admitted with the diagnosis of SCA was conducted at Centegra hospital. Adult patients (>18 years of age) with OHCA/IHCA were included in the study. Information such as demographics, lactate and troponin levels, kidney functions (GFR, S.creatinine), smoking status, previous history of Coronary Artery Disease (CAD), Ejection Fraction (EF) as documented on Echocardiogram, location of SCA (OHCA/IHCA), duration of Cardiopulmonary Resuscitation (CPR), and initial rhythm at the time of CPR was collected. Patients receiving emergent cardiac catheterization (<24 hours post-SCA) were also studied.

Categorical variables were expressed as absolute or relative frequencies and statistically analyzed with Pearson’s Chi-squared test and the Z test for proportions. After assessing distribution normality with KS test, categorical variables with non-normal distribution underwent a logarithmic transformation. When normalization could not be achieved, variables with non-normal distribution were expressed as medians (25th percentile-75th percentile) and were analyzed using U Mann-Whitney analysis. A multiple logistic regression model was constructed in order to estimate odds ratios (CI 95%) for survival status, adjusted by above-mentioned factors. Student’s T-test was used for quantitative variables to evaluate differences between means from two groups (Deceased vs Survivors). Statistical significance was achieved at p <0.05.

Results: The overall survival rate was 34.5% (n=28). Higher lactate levels were noted in deceased in comparison to survivors (5.52±3.55 mmol/L vs 3.95±3.18 mmol/L, p=0.006). Similar results were seen for the longer duration of CPR (16.85±8.61 minutes vs 9.04±5.32 minutes, p<0.001). A statistical significant association was found between final outcome and initial rhythm at the time of CPR $X^2(3, N=81)= 18.23, p<0.001$, smoking status $X^2(1, N=81)= 6.84, p<0.009$, time of intervention $X^2(2, N=37)= 9.87, p<0.007$, and cardiac catheterization results $X^2(2, N=37)= 8.75, p<0.013$. Regression analysis revealed inferior survival outcomes in smokers (OR: 6.752, 95% CI: 1.753-26.006, p=0.006) and patients presenting with PEA (OR: 5.671, 95% CI: 1.474-21.816, p=0.012).

Conclusion: As seen in previous studies, CAD was identified as the most common cause of SCA. Patients receiving early cardiac catheterization showed higher survival rates warranting further long-term studies. Better outcomes were seen in patients presenting with VF. Patients incurring OHCA and requiring prolonged CPR tend to have poorer prognosis. Smoking was found as an independent risk factor associated with increased overall mortality.
EPIDEMIOLOGY OF ATRIAL FIBRILLATION IN PATIENTS WITH CIRRHOSIS: A SYSTEMATIC REVIEW

Introduction: Although Cirrhosis is known to cause structural cardiac changes referred to as Cirrhotic Cardiomyopathy, there is paucity of evidence supporting the association of Atrial Fibrillation (AF) and Cirrhosis. Cirrhosis leads to an enlarged left atrium (LA), high cardiac output and electrophysiological abnormalities which can predispose to arrhythmias. However, the low prevalence of hypertension and routine use of beta-blockers have been postulated to protect against AF. The objective of this study was to conduct a systematic review of the published literature on the epidemiology and impact of AF in patients with Cirrhosis.

Methods: A literature search was conducted using the electronic database engines MEDLINE through PubMed, EMBASE, Ovid, Scopus and Cochrane Library (Cochrane Central Register of Controlled trials and Cochrane Database of Systematic Reviews) from inception to April, 2018 to identify published articles and reports addressing epidemiology of atrial fibrillation in patients with cirrhosis.

Results: The search strategy retrieved 972 papers out of which 3 observational studies were selected with a total of 5232 patients. Out of those 5232 cirrhotic patients (mean age 56.1), 136 (2.6%) were found to have AF compared to 0.95% prevalence in the general population as reported in a large ATRIA study by Alan et al. Only one study reported increased mortality (46%) in Cirrhosis patients with AF compared to control group (39%) but the results could not reach statistical significance. However, we observed a higher prevalence of AF in the younger patients with cirrhosis compared to the overall population.

Conclusion: The results of this systematic review suggest a trend towards a higher prevalence of AF in patients with Cirrhosis. Due to the increasing worldwide prevalence of AF and Cirrhosis, further studies are needed to evaluate their possible association. Moreover, due to a shorter life expectancy and high bleeding risks, further studies are warranted to establish the safety of thromboprophylaxis in patients with Cirrhosis who have AF.
Re-challenging Non-Small Cell Lung Cancer Patients with Immune Checkpoint Inhibitors – A Systematic Review of Safety and Efficacy

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Introduction: Immune checkpoint inhibitors (ICPI) have revolutionized the management of Non-Small Cell Lung Carcinoma (NSCLC), with improvement in overall survival. However, data regarding re-challenging patients with ICPIs who were previously on an ICPI and discontinued treatment is scarce. The aim of this study was to analyze the safety and efficacy of resuming ICPI therapy, particularly inhibitors of programmed cell death receptor and receptor ligand (PD-1/PD-L1) in NSCLC patients who were previously treated with an ICPI agent.

Methods: A systematic search of Medline, Embase, Scopus, and Cochrane Library for studies from January 2010 to August 2018 was performed using controlled vocabulary as well as natural language terms for ICPIs. Studies enrolling NSCLC patients who previously received ICPIs and were re-challenged with PD-1/PD-L1 ICPIs were included. Due to scarcity of data, case reports were also included in our study. Data were compiled from the included studies regarding the response to therapy as well as the reported immune related adverse events (irAEs) attributed to ICPI re-challenge. Infectious adverse events and general patient reported adverse events such as fatigue were excluded as these did not constitute an irAE. Descriptive statistics were employed to report the findings of our systematic review.

Results: Overall, 3426 initially screened studies yielded 3 studies (2 case series and one case report) enrolling 24 patients, of whom 6 (25%) were female. All patients had previously received a PD-1/PD-L1 ICPI. In 23 (96%) patients, the initial ICPI was terminated due to disease progression, whereas treatment was terminated in 1 (4%) patient due to a grade 2 irAE (drug induced alveolitis). Subsequently, 13 (54%) and 11 (46%) patients received pembrolizumab and nivolumab respectively. Of the 24 patients, 5 (21%) patients achieved partial response, 6 (25%) patients had stable disease and 12 (50%) patients experienced disease progression. In 1 (4%) patient, best response could not be evaluated due to early death. A total of 33 irAEs were reported, of which 2 (6%) (interstitial pneumonitis) were grade ≥3 in severity. The most frequently reported irAE of any grade was dermatitis (n=11; 33%), followed by interstitial pneumonitis (n=8; 24%), and diarrhea (n= 7; 21%). No irAE related mortality or treatment discontinuation was reported.

Conclusion: Re-challenging patients with PD-1/PD-L1 ICPI appears to be a safe and viable option in patients with NSCLC. Given the scarcity of data and our small sample size, larger studies in the form of randomized clinical trials are needed to establish efficacy and safety further.

References

ILLINOIS RESEARCH POSTER FINALIST - SUMIT SOHAL, MD

To evaluate the utility of STOP-Bang questionnaire to predict outcomes of rapid response events.

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Introduction: Rapid Response System (RRS) was designed as a safety tool for early detection and intervention of a deteriorating patient on a general floor in a hospital. The patients with Obstructive Sleep Apnea (OSA) including patients at high risk for OSA identified by STOP-Bang questionnaire, have higher rates of RRS events, however their outcomes have rarely been studied. This study is an attempt to identify STOP-Bang score-based risk classification as a predictor of outcome of such RRS events.

Methods: This retrospective cohort study was conducted on patients admitted between November, 2014 and June, 2017 at Presence Saint Francis Hospital. Data was collected by accessing the charts of patients above the age of 18 years, who had STOP-Bang questionnaire filled at the time of admission and had RRS event during the admission. Patients with STOP-Bang scores between 0 and 2 were identified as low-Risk OSA(LR-OSA) and between 3 and 8 as High-Risk OSA(HR-OSA) and outcomes were compared using these groups. Primary outcomes studied was death from any cause or hospice enrollment during the hospital stay. Secondary outcomes included ICU transfer, intubation events, code blue events, duration of mechanical ventilation, duration of ICU stay and duration of hospital stay. Analysis was done with SPSS using chi-square test, Fisher’s exact test and Student’s t test. 2-tailed p value < 0.05 was considered significant.

Results: Out of 519 patients who had RRS events in the study period, 297 patients were eligible for this study. 97(32.66%) patients were identified as LR-OSA and 200(67.34%) were HR-OSA. Patients who were HR-OSA were significantly older than LR-OSA(71.31±15.5 yrs. vs 61.21±22.33 yrs. p<0.0001). Mean STOP-Bang score for LR-OSA was 1.57±0.63 and HR-OSA 3.97±1.19 (p<0.0001). MEWS at the time of RRS and time to RRS from admission was non-significantly lower for HR-OSA vs LR-OSA ((3.86±1.95 vs 4.06±2.10, p=0.426), (76.15±94.14 hrs. vs 82.97±100.14 hrs. p=0.567)). Primary outcome i.e. death or hospice enrollment occurred in 58 (19.53%) patients with non-significantly higher number in HR-OSA vs LR-OSA (38 vs 20, p=0.741). Non-significantly higher number of patients were transferred to ICU(91 vs 37, p=0.230), got intubated(26 vs 12, p=0.879) and had code blue(9 vs 4, p=0.573) in HR-OSA vs LR-OSA groups respectively. Patients with HR-OSA also had non-significant higher duration of mechanical ventilation (0.49±2.04 days vs 0.36±1.46 days, p=0.555), duration of stay in ICU (1.44±2.82 days vs 1.32±2.45 days, p=0.726) and duration of stay in hospital (9.42±8.04 days vs 8.27±5.86 days, p=0.213).

Conclusion: From these results it is apparent that though patients with HR-OSA are older and have more RRS events, their outcomes after RRS do not vary based on their OSA risk assessment including their risk of death or hospice enrollment, transfer or stay in ICU, intubation events, code blue or duration of stay in hospital.
Factors implicated in thoughts of suicide and self-harm among medical students – a single institution’s experience

Authors: Christine Thomas DO, Blake Murphy MS3, William Adams PhD, Brendan Martin PhD, Tania Torres MD, Laura Ozark MD

Introduction: As the medical education community addresses depression and suicidal ideation among students, it is essential to understand factors that positively and negatively affect thoughts of self-harm. The purpose of this study is not only to describe the prevalence of depression and thoughts of suicidality or self-harm among medical students but also to identify risk factors and, most importantly, potential lifestyle factors which may protect students against the development of these thoughts.

Methods: Following IRB approval, an anonymous survey was distributed to all medical students enrolled at Loyola University Stritch School of Medicine between July 2017 and June 2018. Basic demographic information included students’ history of depression or anxiety, responses to the Patient Health Questionnaire (PHQ-9), and questions on regular exercise habits, relationship status, spirituality, communication with family, and access to friends. Major depression was defined as a PHQ-9 score ≥ 10\(^1\), and frequency of self-harm or suicidal ideation was rated on a five-point ordinal scale ranging from not at all (0) to nearly every day (4). Univariable ordinal logistic regression models were used to estimate the odds of more frequent suicidal ideation as a function of students’ demographics. For each model, the proportional odds assumption was assessed using a score statistic\(^2\). Due to survey anonymity, it was not possible to measure paired survey responses; only the first survey response was used for these analyses.

Results: Seven-hundred responses were collected from four medical school class years. Among these responses, 114 (16.3%) were consistent with major depression and 66 (9.4%) reported thoughts of self-harm; 46 of these 66 responses (69.7%) endorsed such thoughts several days of the week, 8 (12.1%) over half the days, and 12 (18.2%) nearly every day. Using students’ first survey response (n=227), a history of depression or anxiety was significantly associated with more frequent suicidal ideation or self-harm (OR=4.46, 95% CI: 1.58 – 12.59; \(p = .005\)), and males were nominally more likely than females to report more frequent thoughts of self-harm (OR = 2.88, 95% CI: 0.97 – 8.56; \(p = .058\)). Conversely, students with a significant other were nominally less likely to report more frequent thoughts of self-harm (OR = 0.33, 95% CI: 0.11 – 0.98; \(p = .047\)). In this sample, there was no association between thoughts of self-harm and class year (\(p = .51\)), regular exercise (\(p = .13\)), access to friends (\(p = .13\)), communication with family (\(p = .10\)), or spirituality (\(p = .45\)).

Conclusion: Our analysis confirms a high prevalence of depression and thoughts of suicide or self-harm among medical students and demonstrates an association between a prior history of depression or anxiety and thoughts of self-harm while also suggesting the presence of a significant other may offer protection against these thoughts.

References

Improving Diabetes Standards of Care and Documentation in a Resident Clinic: A Quality Improvement Project

Authors: Gloria KH Ong, M.D., Amna Anees, M.D., Ancy Jacob, M.D.

Introduction: According to the Centers for Disease Control and Prevention (CDC), 30.3 million Americans are diabetic, making up 9.4% of the United States population. With statistics showing both incidence and prevalence of diabetes diagnoses steadily rising, medical communities must respond to this increasing healthcare burden. Although standards of care have been long studied and published, there are continued barriers to address all quality metrics in a single patient encounter. We present a Quality Improvement project that targets executing and documenting diabetic standards of care in a resident clinic.

Methods: First, patient lists were generated under resident providers that were seen for diabetes evaluations. Pre and post intervention data were gathered from July to September 2017 (n=377), and April to July 2018 (n=533) respectively. For both data sets, we eliminated duplicate, acute care, and non-relevant visits in which diabetes was not thoroughly addressed.

9 standard of care metrics were chosen, including: hemoglobin a1c, urine microalbumin, diabetic foot exam, ophthalmology exam, hepatitis B vaccine, pneumococcal (PPSV23) vaccine, blood pressure control, statin, and aspirin 81 mg use (ASCVD risk>10%). Additionally, we studied whether thorough documentation of such standards of care occurred in each patient chart.

Our intervention was composed of a created template in an electronic medical record system with specific menus to address whether each of the above parameters were completed and documented. Internal Medicine residents were encouraged by faculty to use the template when addressing diabetes for a duration of 4 months (April-July 2018). In addition, notices were placed at each computer workstation, and e-mail alerts were sent to residents to increase awareness.

Results: Analysis of 377 pre-intervention and 457 post-intervention chart reviews were completed using non-paired, one-way Z-tests of two population proportions using a p value of <0.05. We found statistically significant increases in execution of: yearly microalbumin, ophthalmology exam, foot exam, and PPSV23 vaccinations. Although not statistically significant, we did see a clinically relevant increase in Hepatitis B vaccinations.

With regards to metric documentation, we found statistically significant increases in: yearly microalbumin, ophthalmology exam, foot exam, blood pressure control and aspirin.

Overall post-intervention template use was 27%.

Conclusion: With diabetes as the seventh leading cause of death in the United States, immediate action must be taken to meet all the standards of care. Results of this Quality Improvement project displays positive outcomes confirming improved execution and documentation of some quality metrics. Although our template was only used 27%, we have identified barriers to resident compliance. We hope to inspire other resident clinics to implement a comparable tool on various other electronic medical records to achieve similar results. By introducing a simple tool to standardize care, we improved quality and our documentation of one of the leading causes of morbidity and mortality in the United States.
References

Usefulness of a mobile phone application for measurement of respiratory rate in adult patients

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Introduction: Measurement of respiratory rate (RR) is important for the early detection of exacerbation of patients' condition. However, it is sometimes bothersome for healthcare providers to measure respiratory rate visually over 60 seconds (one-minute method). RR measurement using a mobile phone application (app method) has been reported to be accurate and completed in a short time, but investigated only in a pediatric setting. The objective of this study was to validate the performance of the app method for measuring RR compared with the one-minute method in adult patients.

Methods:

Study design: A cross-sectional study

Setting and participants: Nursing school students in a teaching hospital in Japan

Measurements: The movements of the thorax during spontaneous respiration of five adult inpatients were recorded on de-identified videos. Then reference RR was defined by two independent observers. Participants watched these videos and measured the RR with both the app and the one-minute methods. Also, the time taken for the measurement was recorded.

The RR measured by each method was compared with the reference RR. A Bland-Altman analysis was conducted to calculate bias, limits of agreement, percentage error and root mean square error (RMSE). The time taken for the measurement with each method was compared using a t-test.

Results: A total of 59 students participated, with a mean age of 20.9 years. When compared to the reference RR, \( \text{RR}_{\text{app}} \) and \( \text{RR}_{\text{1min}} \) showed a bias of 0.40 breaths per minute (brpm) and 0.65 brpm, limits of agreement of -2.83 to 3.67 brpm and -2.11 to 3.41 brpm, and RMSE of 1.71 brpm and 1.55 brpm, respectively. The mean measurement time with the App method was 22.8 sec, which was significantly shorter than the 65.8 sec taken with the one-minute method (\( p < 0.05 \)).

Conclusion: RR can be measured accurately in a shorter time using a mobile phone application in adult patients.
Meta-analysis of calcitonin gene-related peptide monoclonal antibody for chronic migraine: Assessing efficacy and safety

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Introduction: Historically under-investigated relative to its global disease burden, migraine is the world’s third most common disease and second most prevalent primary headache disorder with an estimated global prevalence of ~18%. Nonetheless, effective treatments with high quality of evidence have been limited and side effect-laden. Furthermore, chronic migraine defined as ≥15 headache days per month for at least 3 months with ≥8 being migraines; is characterized by ~2% global prevalence (WHO), greater disability, and even fewer established treatments. Until recently, benefits of calcitonin-gene related peptide (CGRP) antibodies have not been validated in chronic migraine.

Methods: The PubMed database was searched from inception until present. Various iterations of keyword and MeSH term searches including the terms “CGRP”, “monoclonal antibodies”, and “chronic migraine” were performed. Further RCTs were surveyed from the references sections of all identified studies and meta-analyses. Selected studies investigated specific CGRP monoclonal antibodies vs. placebo in the chronic migraine population for a 12 week time period and reported on >50% reduction in average monthly headache days as a primary or secondary outcome.

Results: Three high quality randomized clinical trials evaluating fremanezumab and erenumab CGRP monoclonal antibodies in 2,038 subjects were included. Outcomes of this meta-analysis showed that CGRP monoclonal antibodies for preventive treatment of chronic migraine significantly increased the >50% reduction in average monthly headache days amongst both higher (RR=1.95 [1.34;2.85]) and lower (RR=1.85 [1.35;2.53]) cumulative CGRP dose groups as compared to placebo. Importantly, an analysis of higher vs. lower cumulative CGRP dose showed no statistically significant difference in attaining >50% reduction in average monthly headache days (RR=1.06 [0.93;1.21]) and carried no heterogeneity (I²=0% [0%;0%]). As for adverse events, those occurring in >2% of patients were more common in the CGRP group (RR=1.27 [0.92;1.77]), however this showed substantial heterogeneity (I²=50% [0%;80%]), whereas serious adverse events were less common in the CGRP group (0.83 [0.46;1.51]) and showed little heterogeneity (I²=0% [0%;47%]). Neither of these trends where statistically significant however, insinuating that side effects were comparable across CGRP and placebo groups respectively.

Conclusion: The CGRP monoclonal antibodies fremanezumab and erenumab studied in chronic migraine to date were equally efficacious in achieving >50% reduction in average monthly headache days at both higher and lower cumulative doses and had fairly similar side effect profiles as compared to placebo. CGRP monoclonal antibodies present as a valid emerging treatment option for chronic migraine suffers with limited treatment modalities who have either failed or not tolerated alternate agents.

References

The Problematic Nature of Fibromyalgia Diagnosis in the Community: Observational Epidemiologic Study

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Introduction: Recently some studies suggested that clinical diagnosis of fibromyalgia is inaccurate and does not reflect current definitions. However, this hypothesis has not been tested in the community. We examined whether fibromyalgia was accurately diagnosed in the community and whether diagnosis was biased by sex.

Methods: We surveyed 3276 consecutive patients attending 25 primary care practices in Kansas using a self-report questionnaire that contained the 2016 modification of the American College of Rheumatology diagnostic criteria in order to determine current fibromyalgia status by criteria (CritFM). We also determined whether the patient had a physician’s diagnosis of fibromyalgia (MDFM), the level of fibromyalgia symptom severity as measured by the polysymptomatic distress scale (PSD), time from MDFM diagnosis, and the use of fibromyalgia pharmacotherapy.

Results: The prevalence of physician (MDFM) and criteria (CritFM) diagnosed fibromyalgia was 6.1% (95% CI 5.3%, 6.9%) and 5.5% (95% CI 4.8%, 6.3%), respectively. However, only 32.2% with MDFM met 2016 criteria (CritFM), and only 35.4% with CritFM also had MDFM. The kappa statistic for diagnostic agreement was 0.296 (minimal agreement). The mean PSD score was 12.4 and 18.4 in MDFM and CritFM, and generalized pain was present in 39.7% and 100%, respectively. The odds ratio for being a woman compared to being a man was 3.2 (95% CI 2.2, 4.9) for MDFM versus 1.9 (95% CI 1.4, 2.8) for CritFM, p = 0.023. Treatment was related to PSD score in MDFM, even in those who did not meet criteria; 68.3% of patients with MDFM received specific fibromyalgia pharmacotherapy.

Conclusion: There is little agreement between MDFM and CritFM. Only 1/3 of MDFM satisfy fibromyalgia criteria, and only 1/3 of patients who meet criteria have a clinical diagnosis of fibromyalgia. Physician diagnosis compared with CritFM is biased and more likely in women. Fibromyalgia treatment is common in MDFM (70.7%) regardless of criteria status. Overall, diagnosis of fibromyalgia by physicians appears idiosyncratic and unrelated to FM criteria. There appears to be no common definition of fibromyalgia in the community.

References

Effects of Gastric Neuromodulation on Crohn’s Disease in Patients with Co-existing Symptoms of Gastroparesis

Authors: Bhatti, Sundus 1; Jaafar, Imad 1; Hassan, Hamza 1; Atassi, Hadi 1; Stocker, Abigail 1; Hughes, Michael 1; Christina Pinkston 1; Dryden Gerald 1; Abell, Thomas 1.

Introduction: Crohn’s Disease (CD) results from chronic inflammation of the gastrointestinal (GI) tract involving TNF-α release. Gastrointestinal electrical stimulation, (GES), a form of neuromodulation used to treat upper GI motility symptoms (UGI Sx), exerts an anti-inflammatory effect via TNF-α suppression. We hypothesized patients with Crohn’s Disease (CD) and UGI Sx may respond to GES.

Methods: We examined 284 patients with gastroparesis (Gp) Sx who underwent GES placement. Patients with Gp Sx were evaluated by a validated GI Sx patient reported outcome. Scores were obtained at baseline, after temporary GES placement and after permanent GES placement. Eleven patients with coexisting CD were analyzed for improvement in their CD Activity Index using the Harvey Bradshaw Index (HBI). A 3-point decrease in HBI indicated a clinical response and an HBI of <5 clinical remission after GES. Data analysis was done using an unadjusted repeated measures analysis, statistically significant if p≤0.05.

Results: Our cohort prevalence of CD was 3.9% (2 M & 9 F, mean age 49.8 yrs). Within the CD & Gp subgroup UGI Sx showed statistically significant improvement after temporary and permanent GES. Within the CD subgroup, 64% of patients showed a clinical response and one patient achieved clinical remission, t score <0.01. CD medications were reviewed before and after GES placement, any changes made do not appear to explain the improvement HBI scores.

Conclusion: CD & Gp patients responded well to GES. Both the interaction of gastroparesis and CD, and the beneficial effects of neuromodulation on CD symptoms warrant additional investigation.
Maryland Research Poster Finalist - Rachit Vakil, MD, MPH

The Association of Baseline Elevation at Various ST Points with Mortality in the Multi-Ethnic Study of Atherosclerosis and Atherosclerosis Risk in Communities Study Cohorts

Authors: Rachit M. Vakil, MD, MPH, David Tian, MD, Yiyi Zhang, PhD, Eliseo Guallar, MD, DrPH, Elsayed Z. Soliman, MD, MSc, MS, Susan R. Heckbert, MD, PhD, Gordon Tomaselli, MD, Wendy Post, MD, MS, David A. Bluemke, MD, PhD, Leonard Ilkhanoff, MD, MS, Joao Lima, MD, Moyses Szklo, MD, DrPH, Saman Nazarian, MD, PhD

Introduction: Prior studies suggest that ST elevation at the J-point is associated with elevated risk of death. We sought to examine the prevalence and prognostic importance of elevation at various ST points in a large multi-ethnic population.

Methods: After confirming data harmonization, we combined ECG, demographics, and mortality data for 19,742 participants from the Atherosclerosis Risk in Communities Study and the Multi-ethnic Study of Atherosclerosis population-based cohorts. Participants were stratified by the presence of ≥ 1 mm inferior (0.5%), lateral (27.8%), inferior or lateral (29.3%), and inferior and lateral (0.2%) ST elevation (at the J-point, mid-point, 60ms after the J-point, and end-point). We utilized Cox proportional-hazards models adjusted for age, gender, ethnicity, source cohort, BMI, education, heart rate, hypertension, left ventricular hypertrophy, smoking status, diabetes, LDL, HDL, and aspirin and/or statin therapy.

Results: The average age at baseline was 56.1 years and 56.2% of the participants were female. Inferior ST elevation at any ST point was associated with increased mortality (HR 1.94, 95%CI 1.32 - 2.84). In contrast, lateral elevation at any ST point was associated with decreased mortality (HR 0.88, 95%CI 0.81 – 0.95). ST-end elevation was more common and drove the association of lateral elevation with decreased mortality. The magnitude of association between inferior ST elevation and increased mortality was strongest when occurred at the mid-ST or J-points. Although the prevalence of elevation varied among subgroups, no additive or multiplicative interactions were noted with gender or ethnicity.

Conclusion: We found that asymptomatic inferior lead ST elevation is uncommon and is associated with elevated risk of mortality regardless of ethnicity. In contrast, asymptomatic lateral ST elevation at the ST-end point is common and is associated with lower risk of mortality compared to participants without ST elevation.
Impact of Hepatitis C Remission on Glycemic Control in Patients with Type 2 Diabetes Mellitus

Authors: Kartikeya Tripathi, MD, Himmat Grewal, MD, Nitin Trivedi, MD, DM, George M. Abraham, MD, MPH

Introduction: Chronic hepatitis C is one of the major causes of chronic liver disease and cirrhosis worldwide. The occurrence of HCV often worsens the glycemic control in patients with preexisting type 2 diabetes mellitus (DM). Although not fully understood, HCV proteins may increase insulin resistance by phosphorylation of serine and threonine residues of the insulin receptors. We aimed to investigate if achieving sustained virological response (SVR) after successful treatment of chronic HCV infection with direct-acting antivirals (DAAs) improves glycemic control.

Methods: We performed a retrospective chart review of patients with chronic hepatitis C and type 2 DM who achieved SVR using DAAs. Baseline demographics and disease characteristics were recorded including age, gender, time of diagnosis of HCV infection, Vibration-Controlled Transient Elastography (Fibroscan®) staging before and after treatment, type, duration and complications of diabetes mellitus (DM), HbA1c before and after achieving SVR, baseline glycemic control and change in anti-diabetic medications. In addition, change in body weight, smoking status, physical activity, and other medications were recorded.

Results: Of the 180 patients with chronic hepatitis C in the study period, 12 had type 2 DM. Eight out of 12 patients achieved SVR. All patients were men with a mean age of 58.4 years. Eighty-seven percent of these included patients had chronic hepatitis C for over ten years. Fibroscan® performed prior to initiation of treatment showed stage F4 in 75 percent, F3 in 12.5 percent and F0 in 12.5 percent of patients. All 12 patients had DM for more than ten years with at least one micro- or macrovascular complication. The mean HbA1c decreased from 9.4 to 6.1 percent after achieving SVR. Thirty-seven percent of patients had a significant reduction in the dose of anti-diabetic medications following SVR with one patient going off all anti-hyperglycemic medications. There was glycemic improvement across all patients without any clinically documented hypoglycemic events. There was no significant change in body weight.

Conclusion: We observed a significant decrease in HbA1c in patients with SVR. This effect was unlikely due to direct effect of DAAs as the response was sustained after the therapy was completed. Although large randomized control studies are required to firmly establish this effect, the findings of our single center study support prompt DAAs treatment for HCV especially in patients with type 2 diabetes mellitus. Better glycemic control is likely to delay or prevent short and long-term complications of diabetes.
Quality improvement project for prevention of infusion related reactions of daratumumab therapy in multiple myeloma patients

Authors: Vishal Jindal, MD, David Scott Shepro, MD, Leigh Pranckevicius, Ahmad Daniyal Siddiqui, MD, Department of Medicine, St. Vincent Hospital, Worcester, MA

Introduction: Daratumumab (DARA) is an IgG1K human monoclonal antibody that binds to CD38 and inhibits the growth of myeloma cells. The most common adverse reaction of DARA is infusion related reactions (IRRs). To prevent IRRs, pre-infusion and post-infusion pre-medications are recommended. However, 45% of patients still develop infusion reactions as per previous studies. Most of the IRRs occur in the first four hours, with a median of 1.4 hours and especially after increasing the infusion rate from 50ml/hr to 100 ml/hr at 1hr. Three patients who received DARA with standard protocol all had infusion reactions before 2 hours. This led to introduction of a quality improvement project to give pre-medications at 1hr along with usual pre-infusion and post-infusion medications.

Methods: This is a quality improvement project with a goal to prevent IRRs with standard therapy plus pre-medications at one hour of the first two infusions. Standard therapy includes pre-infusion medications (diphenhydramine 50mg IV, methylprednisolone 100mg IV, famotidine 20mg IV, cetirizine 10mg and acetaminophen 975 mg) and post-infusion medications (20mg oral prednisone on day 2 and 3). In this project a total 8 patients were treated with pre-medications (methylprednisolone 40mg IV, famotidine 20mg IV, and diphenhydramine 25mg IV) at one hour, after the initiation of DARA infusion along with standard therapy. IRRs were graded as per CTCAE 4.03. Results were compared with the previous studies which used standard timing of therapy.

Results: Out of 8 patients treated during their initial infusions of DARA, one patient (12.5%) developed grade 2 IRRs. The other 7 patients experienced no IRRs. The incidence of infusion modification due to IRR was 12.5 % in our study as compared to 42% with standard pre-medications schedule. There have been no short term or long term adverse effects seen with this change in our pre-medications schedule.

Conclusion: With the addition of pre-medications at 1 hour after initiation of DARA infusion, the rate of IRRs reduced dramatically. This has led to a decrease in the total infusion time and increased the tolerability. Additional clinical trials suggested to confirm this outcome.
Surgical Left Atrial Appendage Occlusion During Cardiac Surgery: A Meta-analysis

Introduction: The left atrial appendage is a common site for intracardiac thrombus formation in patients with atrial fibrillation (AF). Surgical left atrial appendage occlusion (s-LAAO) during concomitant cardiac surgery has been evaluated as an effective treatment approach to reduce the risk of stroke and embolic events.

Methods: We performed a comprehensive literature search through May 1st 2018 for all eligible studies comparing s-LAAO versus no occlusion in patients undergoing cardiac surgery. Clinical outcomes during follow-up included: embolic events, stroke, all-cause mortality, atrial fibrillation, reoperation for bleeding and postoperative complications. We further stratified the analysis based on propensity matched studies and AF predominance.

Results: Twelve studies (N=40,107) met the inclusion criteria. 13,535 patients received s-LAAO during cardiac surgery while the remaining 26,572 patients did not receive s-LAAO. The mean (SD) age of the study population ranged from 50.7 (12.4) years to 77.4 (6.8) years. The follow-up period ranged from in-hospital only to 109.2 months. s-LAAO was associated with lower risk of embolic events (OR: 0.63, 95% CI: 0.53 to 0.76; p< 0.001) and stroke (OR: 0.68, 95% CI: 0.57 to 0.82, p< 0.0001). Stratified analysis demonstrated this association was more prominent in the AF predominant strata. There was no significant difference in the risk of all-cause mortality (OR: 0.83, 95%CI: 0.51 to 1.36, p= 0.46), followup atrial fibrillation (OR: 1.41, 95% CI: 0.79 to 2.52, p= 0.24), reoperation for bleeding OR: 0.98, 95% CI: 0.57 to 1.69, p= 0.94) and postoperative complications (OR: 1.44, 95% CI: 0.91 to 2.25; p= 0.12).

Conclusion: Concomitant s-LAAO during cardiac surgery was associated with lower risk of follow-up thromboembolic events and stroke, especially in those with AF without significant increase in adverse events. Further randomized trials to evaluate long-term benefits of s-LAAO are warranted.

References

“All Eyes on You”: A Covert Observational Study on Contact Precaution Compliance in Six Hospitals at the Detroit Medical Center.

Aim 1: To covertly monitor the overall compliance of contact precautions (CP) among healthcare providers during routine patient care;

Aim 2: To analyze and compare the compliance rates of individual CP (hand hygiene, glove and gown) components among HCW during the care of patients under

Methods: A prospective observational study was conducted from July 2017 to February 2018 in six hospitals affiliated with Detroit Medical Center (DMC). Trained anonymous observers performed the compliance audits for CP. Standardized definitions and monitoring tool (speedy audit app) were used to record these data. Components of CP recorded were (1) HH before donning (2) proper gowning and gloving techniques upon entering the patients room, (3) doffing the gown and gloves properly after leaving the room and (4) HH after doffing. A pilot study providing targeted education focused on strict adherence of HH practice before donning gloves was implemented in one hospital.

Results: A total of 6274 observations were done. The overall CP bundle compliance was 38%. The most common type of HCW observed included Nurses (registered nurse and nursing student) - 46.8%; Physicians (attending’s, residents, fellows) - 28.4%; Service workers (SWS) including PCA, Environmental Service, Food service, Patient transporter, Social worker, Pastoral care - 14%; Allied Health Professions(AHP) including Dietician, Blood Collection, Physiotherapist, Radiology Tech, Respiratory Therapist-3.7%; Medical students (MS)-1.3%; Unknown profession-5.7%. Overall CP bundle compliance rate among HCW was as follows: Nurse-44%, Physician-42%, AHP-30%, SWS-21%, Unknown-13%. Overall individual CP compliance were 49.1% for HH, 79.9% gloving, 62.3% gowning. HH compliance before donning were strikingly low (40%) compared to the compliance after doffing (62%). The HH compliance before donning was relatively low among AHP (30.3%) and SWS (22.6%) when compared to nurses (48.4%), MS (47.9%), physicians (41.4%). Within a month of the targeted education initiative at the pilot hospital we noticed a drastic increase in the compliance rates with HH practice before donning gloves was implemented in one hospital.

Conclusion: The overall compliance of CP bundle was low among all HCWs. The low HH compliance before donning the gloves can be linked to the common misconception that gloves are a substitute to hand hygiene. Recognition of this knowledge gap and targeted education on adherence to HH before donning gloves has led to improved compliance rate among HCWs at DMC.
Long-Term Follow-Up of Patients With Macroprolactinomas

Introduction: Prolactinomas are the most frequent functioning pituitary adenomas. Management usually consists of medical treatment with dopamine agonists with or without surgery. Limited data exists on long-term follow-up of patients with large prolactin secreting tumors treated with medical therapy. The aim of our study is to describe the clinical presentation of large prolactinomas, patient characteristics and response to medical therapy over time.

Methods: This was a retrospective chart review of patients seen and followed in the outpatient setting. Data was collected regarding age at diagnosis, sex, initial tumor size, presenting symptoms/hormone abnormalities and radiologic/hormonal response to therapy.

Results: Thirteen patients were included in the analysis. Ages ranged from 23-78, eleven were men and two were women. The length of follow-up ranged from 2-22 years. The most frequent presenting symptoms were impotence in men and amenorrhea/galactorrhea in women. All patients had elevated prolactin levels (355-10,000 ng/dl) and large pituitary lesions (1.4-4.8cm). Every male patient had low testosterone levels at presentation. All patients received therapy with dopamine agonists. Significant clinical, biochemical and radiologic responses were observed. Prolactin levels returned to virtually normal in all patients and testosterone levels recovered in most male patients. Majority of patients continue maintenance therapy with Bromocriptine or Cabergoline, however, some have no evidence of tumor recurrence after medication discontinuation.

Conclusion: Our study confirms that dopaminergic agents are the agents of choice for patients with macroprolactinomas. These medications are safe and result in significant reduction in tumor size, near normalization in prolactin levels and significant clinical improvement.
Improving Transition of Care by Increasing Post-Hospital Discharge Follow-up Rates at the Internal Medicine Residency Clinic

Authors: Hafiz Khan, Mahin Khan, Siddique Chaudhary, Ahsan Wahab, Bilal Nasir, Juan Gonzalez, Farah Al-Sabie, Shagufta Ali

Introduction: Transition of care (TOC) is defined as the movement of the patient from one setting of care to another. TOC can create gaps in the quality and safety of patient care and result in readmissions and medical errors exerting a burden on the healthcare system. Studies investigating the difference in post-discharge follow-up (FU) have shown lesser FU rates within 7-14 days due to various reasons. In addition, the patient with resident PCP had higher (20%-25%) odds of readmission compared to faculty primary care.

The ACGME also considers TOC as one of the milestones for the competency evaluation of the residents. Moreover, CMS has now tied reimbursement to TOC visit within 7-14 days. We had no TOC concrete system in place to confirm FU appointments for the patients at the time of discharge from the hospital.

Methods: CQI AIM: Our project aim was to improve the 2-week post-discharge outpatient FU rates for patients at the Internal Medicine Residency Clinic (IMRC).

The study period was from January – August 2018. After obtaining IRB and RAB approval, baseline data were collected on following inclusion criteria: All patients who have an established PCP (resident Physician) in IMRC or they choose the resident physician as their new PCP at the time of discharge from the hospital. We identified the stakeholders and met as an interdisciplinary TOC team every month. We did process mapping and root cause analysis for timely TOC obstacles. We educated the residents, asked for input and shared root-cause analysis during the monthly quality improvement forum. CQI interventions were planned and implemented as part of the new TOC policy. We re-collected and analyzed data after 1, 3 and 6 month.

Results: Pre-intervention data showed that 17.6% the patients were for TOC within 14 days after discharge, 35.29% after 14 days and 41.17% did not make an appointment after discharge. Insurance obstacles, improper telephonic communication, post-discharge coaching, and care plan development were found to be the most important factors for poor FU rates.

Post-intervention data were reviewed at 1, 3 and 6 months after the implementation of TOC policy. At one month, the total number of inpatient admissions was 40; out of which 27.5% had FU within 7 days, 2.5% within 14 days and 57.5% had no FU. 5% of patients were sent to the rehab facility and 7.5% passed away. FU rates within 14 days were further increased to 55.3% and 48.7% at 3 and 6 months. Non-compliance and insurance were major limiting factors for patients who could not do schedule FU appointment. Overall, FU rates were significantly increased from 17.6% to an average of 53.5% over the span of 6 months.

Conclusion: TOC is one factor, which if done successfully can improve patient care, provide efficient continuity of care, and reduce readmission rates. Our data suggest that by educating the residents and implementing a standardized process at the time of discharge, we can improve the TOC visits in a timely fashion.

References

Concomitant Use of Direct Oral Anticoagulants and Aspirin for Cardiovascular Disease Prevention in Atrial Fibrillation and Flutter

Authors: Ahmad Said, M.D, Scott Keeney, D.O, Alexandra Halalau, M.D., F.A.C.P

Introduction: Atrial fibrillation (AF) affects approximately 3% of adults in the USA\(^1\), and is associated with an almost two-fold increased mortality\(^2\). Previous reviews showed no benefit in terms of major adverse cardiovascular events (MACE) and bleeding rates when oral anticoagulants were combined with antiplatelets in AF\(^3\).\(^4\).\(^5\). Direct oral anticoagulants (DOACs) are increasingly used in thromboembolic disease prevention in patients with AF and atrial flutter (AFL). We hypothesize that the concurrent use of DOACs and aspirin in those subjects will result in less cardiovascular events compared to DOAC therapy alone.

Methods: This was an observational retrospective study that included adults with nonvalvular AF and AFL on a DOAC between 2010 and 2015 in the Beaumont Health System. The population was classified into two study groups based on the presence or absence of concurrent aspirin use. Subjects taking other antiplatelet agents or using anticoagulants for indications different than AF or AFL were excluded. The primary outcome was MACE, defined as acute coronary syndromes, ischemic stroke, and systemic embolism. Secondary outcomes were bleeding and death. A minimum of two years of follow up was used to identify outcomes. Propensity scores were calculated for baseline characteristics and used to achieve balance between the two groups. Hazard ratios and 95% confidence intervals were obtained using a Cox proportional hazards model for all outcomes.

Results: 3,817 subjects were on a DOAC alone and 3,638 were taking a DOAC with aspirin (combination group). 56% of the patient population was male, and 80% Caucasian. Both groups had similar prevalence of heart failure, diabetes, stroke, coronary disease, COPD, GI bleed, kidney disease, and peptic ulcer disease. Propensity scores were used to weigh rates of tobacco use, hypertension, CHADS2-VASc scores, and certain medications such as beta-blockers, angiotensin converting enzyme inhibitors, statins, non-steroidal anti-inflammatory drugs and proton pump inhibitors. More MACE and bleeding events occurred in the combination group with a hazard ratio of 2.12, 95% confidence interval [1.85,2.43] and 1.31, 95% confidence interval [1.17,1.46] respectively.

Conclusion: Using a large sample of subjects with AF and AFL, our study compared the rates of MACE and bleeding events among individuals receiving DOAC alone versus DOAC with aspirin, and demonstrated an increase in MACE as well as bleeding events when receiving combination therapy. These results align with a meta-analysis from 2016 that compared MACE and bleeding between oral anticoagulants with and without antiplatelets\(^3\). Based on this evidence, we recommend against combining DOACs with aspirin as a thromboembolic prevention strategy in AF or AFL; however, this is a retrospective study that did not adjudicate medical histories and outcomes; hence, randomized controlled trials are necessary to eliminate confounding and establish concrete guidelines.

References

Every Visit Counts; Rollout of a Vaccination Program in Resident Clinic During a Hepatitis A Virus Outbreak in Michigan: a Quality Improvement Study

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Introduction: Hepatitis A virus (HAV) infection is a vaccine-preventable illness. In 2016, national HAV vaccine (HAVV) uptake (2 doses) was 9.5% for adults, with only 13% of adults having immunity to HAV in Michigan. In the last two years Michigan has experienced the largest outbreak in U.S. history with over 900 cases of HAV infection and 28 deaths. The primary objective of this study is to determine the feasibility and sustainability of a vaccination program to increase the uptake of vaccines during an outbreak.

Methods: This is an ongoing quality improvement (QI) project in the outpatient resident clinic at Beaumont Hospital, Royal Oak, the largest outpatient clinic in Southeast Michigan that serves the communities most affected by the Michigan outbreak. The entire clinic staff (registration, nursing, pharmacy, residents, and attendings) was engaged in developing a feasible and sustainable QI intervention. Fliers with information about HAV were given to the patients and medicine residents. A questionnaire for patients to determine eligibility and willingness to receive the vaccine was systematically administered to each patient that visited the clinic beginning February 12, 2018.

Results: From February 12th - November 25th, a total of 1442 questionnaires were administered and 505 HAVV were given. We analyzed a subset of patients who received the questionnaire between February 12th - April 29th and followed them until November 25th (n=567). Of these, 183 (32.3%) patients received a dose of HAVV at initial or subsequent visit. Of those that received HAVV at initial visit (137/567, 24.2%), 58.4% were females, 30.7% had diabetes, 21.1% with substance use, and 3.6% with HIV. Of note, although 50/137 (36.5%) patients did not answer "Yes" on the questionnaire to request HAVV, they subsequently received it after discussion with their provider. In the month prior to initiating the quality improvement project, only 4 patients received HAVV.

Conclusion: In less than a year from initiation of our intervention, we demonstrated that providing direct education to patients at the time of their clinic visit significantly increased uptake of vaccination. In our experience, these patients would otherwise only have been vaccinated as part of postexposure prophylaxis. Challenges and barriers to increasing vaccine uptake included uncertainty regarding cost of HAVV, physicians forgetting to order the vaccine, physicians not seeing the questionnaire in patient folders, and referral of Medicare patients to the health department or an outside pharmacy due to cost. Overall, this could serve as a model to increase uptake of other adult immunizations as a public health measure.
National Trends and Outcomes of Non-autoimmune Hemolytic anemia in Patients with Alcoholic Liver Disease: Results from the Nationwide Inpatient Sample

Authors: Tooba Tariq, MD (1); Matthew Masaru Mayeda BS, MPH (2); Austin Joseph Parsons BS, MS (2); Furqan Irfan MBBS (MD), PhD (2), 1. Department of Internal Medicine, Western Michigan University Homer Stryker MD School of Medicine, 2. Michigan State University, College of Osteopathic Medicine

Introduction: Alcoholic liver disease is associated with certain chemical abnormalities which can result in structural and metabolic alterations in the RBC membrane leading to premature destruction of erythrocytes and hemolytic anemia of varying severity. Prevalence and inpatient burden of alcoholic liver disease has increased over past several years however little is known about similar trends of hemolytic anemia (non-autoimmune) in this patient population. This study was done to identify the burden of non-autoimmune hemolytic anemia in hospitalized patients with co-existing alcoholic liver disease and to identify patient related factors and the hospitalization outcomes.

Methods: The Nationwide Inpatient Sample (NIS) database was used to extract data from 2012 to 2014, for patients admitted with a primary diagnosis of non-autoimmune haemolytic anemia and co-existing alcoholic liver disease. International Classification of Diseases (ICD-9-CM) were used to identify patients; Non-autoimmune hemolytic anemia, unspecified (ICD-9-CM: 283.10), Other non-autoimmune hemolytic anemias (ICD-9-CM: 283.19), acute alcoholic hepatitis (ICD-9-CM: 571.1), alcoholic cirrhosis of liver (ICD-9-CM: 571.2), and alcoholic liver damage, unspecified (ICD-9-CM: 571.3). The 2012-2014 NIS data was utilized to study the incidence of hospitalization admissions, patient demographics, hospital factors, inpatient charges and length of hospitalization.

Results: A total of 189,280 hospitalizations were identified with diagnosis of Non-autoimmune haemolytic anemia in patients with alcoholic liver disease for years, 2012-2014. Majority of the patients were males (69.4%) with mean age of 52.0 years. The rate of hospitalisations increased from 19.7 per 100,000 in 2012, to 20.3 per 100,000 in 2014, (p =0.007; 95% CI; -0.0124 to 0.0135) . Patients presented in far greater numbers to urban teaching hospitals (n=111605, 59.0%) and urban non-teaching hospitals (n=64240, 33.9%) compared to rural hospitals (n=13435, 7.1%). The regional distribution of the hospitalizations were the following: Southern region (n=73730, 39.0%), Western region (n=44100, 23.3%), Midwest region (n=37220, 19.7%) and Northeast region (n=34230, 18.0%). The mean length of hospital stay remained approximately 6 days throughout the study period. However, the mean inpatient charges of those admitted increased from $50,400.79 in 2012, to $55,870.77 in 2014 (increase of 9.8%).

Conclusion: It is imperative to recognize the importance of hemolytic anemia in alcoholic liver disease since it might be more common than initially thought as seen by increasing rates of hospitalizations. Understanding the burden can lead to timely recognition which can prevent unnecessary diagnostic or therapeutic interventions which are associated with increased healthcare costs. For instance, in the setting of alcoholic hepatitis, calculation of discriminant function may be misleading (since much of the bilirubin may be from hemolysis rather than liver inflammation) which may change the decision for initiation of glucocorticoids.
Can Internal Medicine Residents Accurately Detect Inpatient Lower Extremity Deep Venous Thrombosis with Compression Ultrasonography?

Authors: Matthew C. Wilkins, MD: Renny Abraham, MD; Sima Salahie, MD; Mehmet Alpas, MD; Lamia Aljundi, MD; William Dillon, MD; Philip Vendittelli, MD; Irfan Majeed, MD; Fawad Shahid, MD; Abdelkader Chaar, MD; Raghavendra Kamath, MD.

Introduction: Deep venous thrombosis (DVT) is a common disorder associated with significant morbidity and mortality. Formal venous duplex ultrasonography (FVDUS) is limited by frequent lack of 24 hour availability. Point-of-care ultrasonography (POCUS) is a rapidly developing skill. Despite this growing application of POCUS in various disciplines, a similar commitment has yet to be established within the internal medicine community. Current literature demonstrated a considerable diagnostic accuracy of acute DVT by emergency department physicians. We propose that internal medicine residents with limited ultrasonography skills can accurately detect inpatient DVTs with similar results.

Methods: Single center, prospective study examining the use of Proximal-Leg Compression Ultrasonography (PL-CUS) examination by novice internal medicine residents. Residents received 30-45 minute didactic session in addition to POCUS video lecture. Participants consisted of patients admitted under the care of internal medicine residents with concerns for DVT warranting FVDUS. Compressibility of the common femoral, femoral and popliteal vein were examined.

Results: 24 PL-CUS were compared with FVDUS. Prevalence of DVT was 20.8%. PL-CUS studies yielded a sensitivity of 80% and a specificity of 94.7% with a diagnostic accuracy of 91.7%. PL-CUS examination varied from 5-30 minutes. Median time delay between ordering FVDUS and FVDUS preliminary results was 15:02 hours.

Conclusion: Our study showed that PL-CUS performed by internal medicine residents with limited training can yield quick and accurate results. Thus allowing prompt initiation of therapy and avoiding unnecessary therapies and delay in discharges.
Battling the Opioid Epidemic: Minimizing Risk with Naloxone Intranasal Spray

Authors: Carleigh Zahn DO, Susan Bannon MD, Lauren Lamie DO, and David Lee MS

Introduction: Opioid overdoses have become endemic in the United States. With any opioid use there is a risk for overdose, accidental or intentional. Over the past 2 decades, medical providers have increased opioid prescribing for non-cancer pain - this now represents half of the opioids dispensed in the United States. Furthermore, over the past decade, opioid overdose has surpassed motor vehicle accidents as the primary cause of accidental injury death in the United States. Naloxone, an opioid antagonist that displaces mu receptor agonists, counters respiratory depression brought on by opioid overdose. It is an especially promising overdose tool as family, friends, or bystanders can administer it providing reversal of the effects of most opioid overdoses.

Methods: Our objective was to make intranasal naloxone prescriptions available to patients prescribed chronic opioids as part of the new clinic standard of practice.

Inclusion criteria included age greater than or equal to 18 years of age, current patient of the Western Michigan University Homer Stryker M.D. School of Medicine Internal Medicine or Medicine/Pediatrics clinic, and currently receiving chronic opioid therapy: defined as a patient receiving and utilizing opioids most days of the month for greater than 3 months. Exclusion criteria included pregnant patients, those on hospice, or those actively being treated for malignancy. In total, 88 patients fulfilled the criteria.

Opioid overdose patient information packets were mailed to these patients. The informational packets included a letter of explanation of the quality improvement project and its aims, the “Narcan Nasal Spray Quick Start Guide” (used with permission from Adapt Pharma), information on what an opioid overdose is/may appear like, contact information so that the patient could reach out if they had questions/concerns, and an intranasal naloxone prescription.

Western Michigan University Homer Stryker M.D. School of Medicine IRB review committee approved this as an exempt study.

Results: Initial patient responses were mixed. However, this opened up avenues of communication regarding the risks of opioid use. This increase in communication and education had an overall positive impact in our clinic according to patients and residents. Part two of this project is currently underway and examines alternative educational opportunities as well as quantification of the total number of naloxone prescriptions filled at our clinic pharmacy pre and post-intervention.

Conclusion: Studies contend that naloxone rescue kits are effective in reducing opioid overdose rates. With reassurance from such studies, post-intervention efforts to “saturate” at-risk communities with naloxone have accelerated. As a result, we felt that implementing an opioid reversal standard in our clinic was important for our patients. Our study results agree that instigating naloxone has proven beneficial in opening communication pathways and safety measures for our patient population.
Extended Duration of Thromboprophylaxis for Medically Ill Patients: A Meta-analysis of Randomized Controlled Trials

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Introduction: Patients hospitalized for an acute medical illness are at increased risk of venous-thromboembolism (VTE). The benefit of extended duration thromboprophylaxis in these patients beyond hospital remains controversial. Therefore, we performed a meta-analysis of randomized controlled trial (RCTs) examining the efficacy and safety of extended-duration anticoagulation prophylaxis for VTE prevention in this high-risk population.

Methods: An electronic database search was conducted utilizing PubMed, Embase and Cochrane Library to include all randomized controlled trials (RCTs) comparing between extended-duration versus short-duration thromboprophylaxis in medically ill patients with high risk of VTE. The primary efficacy outcome was the composite events of asymptomatic deep venous thrombosis (DVT), symptomatic VTE (including symptomatic proximal DVT and non-fatal pulmonary embolism), and death from VTE-related causes. The primary safety outcome was major bleeding defined as fatal bleeding, critical site bleeding, bleeding with a drop-in hemoglobin of ≥ 2 mg/dl or requiring transfusion of ≥ 2 units. Risk ratio (RRs) and 95% confidence intervals (CIs) were calculated using a random-effects model. Subgroup and meta-regression analyses were performed.

Results: Five RCTs were included totaling 40,124 patients. Mean age 71 years and 50.5% were male. Extended-duration thromboprophylaxis ranged from 28 to 45 days post-discharge. We found that extended-duration thromboprophylaxis was associated with a significant reduction in the primary efficacy outcome compared with standard-duration therapy (RR 0.75; 95% CI 0.67-0.85; P<0.01). Additionally, there were significantly reduced rates of symptomatic VTE and asymptomatic DVT (RR 0.53; 95 % CI 0.33-0.85; P<0.01 and RR 0.81; 95% CI 0.71-0.94; P<0.01). However, there were no significant differences between both groups in VTE-related death (RR 0.81; 95% CI 0.60-1.10; P=0.18) or all-cause death (RR 0.97; 95% CI 0.88-1.08; P=0.64). In contrast, extended-duration thromboprophylaxis was associated with increased risk of major bleeding (RR 2.04; 95% CI 1.42-2.91; P<0.01) as well as non-major clinically relevant bleeding (RR 1.81; 95% CI 1.29-2.53; P<0.01). In subgroup analysis, the primary efficacy outcome was significantly reduced in those aged > 75 years. There was increased trend of increased risk of bleeding in women and patients older than 75 years with extended-duration strategy. In meta-regression analysis, we did not find any modifier effects for primary efficacy outcomes based on study-level covariates including age, duration VTE prophylaxis, body mass index (BMI), proportion of patients who have history of cancer, history of previous DVT, and reason for admission.

Conclusion: Among hospitalized medically ill patients, extended-duration thromboprophylaxis was associated with a decreased risk of composite events of the primary efficacy outcome and symptomatic VTE with no significant difference in all-cause death or VTE-related death. Furthermore, there was a significantly increased risk of major bleeding with extended-duration thromboprophylaxis. Benefits and risks should be weighed before initiating extended-duration thromboprophylaxis until further well-controlled RCTs identify patients with clear benefit and low risk of harm.
Triple versus Dual Inhaler Therapy in Moderate-to-Severe COPD: a Systematic Review and Meta-analysis of Randomized Controlled Trials

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Introduction: The management of chronic obstructive pulmonary disease (COPD) is rapidly evolving, especially with triple-inhaler therapy. We aimed to perform a meta-analysis to ascertain the safety and efficacy of triple-inhaler therapy consisting of an inhaled glucocorticoid (ICS), a long-acting muscarinic antagonist (LAMA), and a long-acting β2 agonist (LABA) when compared with dual-inhalers consisting of ICS/LABA or LAMA/LABA.

Methods: We performed an electronic database search utilizing PubMed, Embase and Cochrane library to include randomized controlled trials (RCTs) comparing between triple and dual-inhaler therapy in patients with moderate-to-severe COPD. Pooled rate ratio (RR) or odds ratio (OR) for dichotomous data and weighted mean difference (MD) for continuous data were calculated with their corresponding 95% confidence interval (CI) using a random effects model. Meta-regression analyses were performed for study-level covariates (mean age, duration of treatment and follow-up, gender, smoking status and baseline forced expiratory value in one second (FEV1) percent of predicted value).

Results: Our study included 12 RCTs totaling 19,322 patients with a mean age of 65±8.2 years and 68.2% were male. Pooled analysis demonstrated a significant reduction in moderate-to-severe COPD exacerbations with triple therapy (RR 0.75; 95% CI 0.69-0.83; P< 0.01) with no significant subgroup difference when triple therapy was compared to ICS/LABA or LABA/LAMA. Meta-regression analyses did not suggest any modifier effects for COPD exacerbations based on study-level covariates. Additionally, trough FEV1 (measured by litres) increased significantly in the triple therapy arm compared to dual therapy (MD 0.09 L; 95% CI 0.07-0.12; P<0.01). In subgroup analysis, triple therapy was associated with a significantly greater increase in trough FEV1 from baseline when compared to ICS/LABA rather than to LABA/LAMA dual therapy with significant subgroup interaction (P value <0.05). In meta-regression analyses, patient with higher baseline FEV1% of predicted value were more likely to benefit from triple-inhaler therapy. Furthermore, triple therapy was associated with a significant reduction in the mean St. George’s Respiratory Questionnaire (SGRQ) score (MD -1.67; 95% CI -2.02 – -1.31; P<0.01), and more patients experienced ≥ 4 points reduction of SGRQ score (OR 1.27; 95% CI 1.19-1.35; P<0.01). There were no significant differences in the incidence of serious adverse events, any adverse events, cardiovascular adverse events, adverse events leading to medication discontinuation, or pneumonia between both groups.

Conclusion: In moderate-to-severe COPD patients, triple therapy was associated with a reduced risk of COPD exacerbations when compared with dual therapy. Additionally, triple therapy was associated with a significant improvement in lung function measured by an increase in the absolute FEV1, and a significant improvement in the quality of life measured by a decreased SGRQ score. There were no significant differences between triple therapy and dual therapy regarding incidence of any adverse events, serious adverse events, pneumonia, or cardiovascular events.
Introduction: For people with end-stage renal disease on a renal replacement therapy (RRT), “travel” and “independence” are rated as 2 of the top 5 factors that inform their choice of a treatment modality. While home dialysis modalities offer patients a high degree of independence, the most common RRT in the USA is in-center hemodialysis (IHD). The limits imposed by IHD treatment can present a variety of challenges for patients who wish to travel. This exploratory study explored how IHD patients managed their travel and the role of dialysis social workers in executing travel arrangements for patients.

Methods: An interview-based, qualitative study was conducted with IHD patients being treated at a large Midwestern Medical Center and community-based dialysis social workers. Data collection was conducted from August 2017 to September 2018. Patients were screened from an inpatient nephrology consult panel and the patients enrolled in the study provided contact information for their dialysis social workers. Interviews with patients focused on experiences before, during and after travel, and social workers were asked to describe their role in helping patients who wish to travel. Interviews were conducted until saturation of themes was reached. Two coders used a grounded theory (constant comparative) approach to analyze the data from verbatim transcriptions. The study protocol was approved by the University’s IRB.

Results: Sixteen patients and eight social workers were enrolled in the study. The patient sample included an equal number of women and men (n=8), 13 whites (81.3%), a mean dialysis vintage of 5.3 years, and an average of 4.4 domestic trips completed. Only 1 patient reported 2 international trips. Social workers were female (100%), the majority were white (n=7; 87.5%), and they reported an average of 2.5 travel requests/month. Preliminary findings for patients indicated that limited knowledge of dialysis facility options and uncertainties about staff at host dialysis units were key concerns in preparing for and engaging in travel. Social workers described insurance literacy and confirming “chair times” as key factors in planning out-of-state travel. There is limited research on travel issues for chronic IHD patients and this exploratory investigation is among the first to articulate barriers and facilitators associated with travel from the perspective of patients and social workers. Guidelines on travel for IHD patients should be readily available and incorporated into ongoing patient education, especially when patients initiate RRT.

Conclusion: This study identified multiple concepts and perspectives surrounding travel arrangements in chronic IHD patients. Promoting and supporting travel for IHD patients can serve to increase their sense of autonomy and provide opportunities to improve their quality of life.
Minnesota Detoxification Scale (MINDS) Assessment Protocol for Treatment of Alcohol Withdrawal, Pre and Post Implementation Comparison.

Authors: Dr. Kristopher Holaday, Dr. Steven Hanovich, Dr. David Beddow, Dr. Love Patel

Introduction: Acute alcohol withdrawal is commonly encountered in general hospital settings and can result in serious consequences. Symptoms and complications of withdrawal represent medical emergencies that carry significant clinical risk and requires attentive medical management. Although there is no national guideline for the assessment and treatment of acute alcohol withdrawal, the current standard of care favors symptom triggered therapy with use of an assessment scoring system (CIWA is the most common) and the drug of choice is the benzodiazepine group. In the spring of 2014, a pilot was implemented at Unity Hospital (part of Allina Health, MN) with the intent of systematizing treatment of alcohol withdrawal at Allina Health using the Minnesota Detoxification Scale (MINDS) assessment for diagnosis with diazepam as a drug of choice to treat acute alcohol withdrawal. By direct comparison MINDS assessment includes fewer screening domains than CIWA and less subjective variation. Treatment for a positive screening utilizes a longer-acting benzodiazepine with a set dosing schedule administered earlier upon recognition of withdrawal symptoms. By the end of 2016, all Allina hospitals had implemented the use of MINDS protocol for patients presenting with alcohol withdrawal symptoms. This study aims to address whether implementation of the MINDS protocol order set for diagnosis and treatment of acute alcohol withdrawal results in a meaningful, measurable improvement in Allina Hospital’s patient outcomes.

Methods: This study will use data from patients who have been treated for alcohol withdrawal at Allina Health hospitals both before implementation of the MINDS protocol and after implementation of the MINDS protocol, between January 2013 and December 2017. The analytic approach will involve comparing the “pre-MINDS” and “post-MINDS” groups of patients with multiple linear and logistic regression analyses. Based on existing data, an estimated 25,000 patients will contribute data to the various analyses of outcomes and complications under this study. A preliminary review of the MINDS alcohol withdrawal protocol at Unity Hospital was conducted using patient data from pre-implementation, 03/01/2013 - 01/26/2014, and post implementation, 03/10/2014 - 11/05/2014. The study’s primary outcome will be hospital length of stay (LOS). Secondary outcomes include: readmissions (ED or all-cause readmission within 30 days), calendar days receiving benzodiazepines and total dose administered, number of ICU stays, number of Green alerts, restraints used, and 1:1 attendant use.

Results: Review of patient data pre-implementation, 03/01/2013 - 01/26/2014, and post implementation, 03/10/2014 - 11/05/2014, of the MINDS protocol at Unity Hospital demonstrated a pre-MINDS LOS of 113.05 hours/4.71 days, and a post-MINDS LOS of 76.5 hours/3.2 days. The average total diazepam equivalents administered pre-MINDS was 102.4 mg, and post-MINDS 87.3 mg.

Conclusion: Preliminary data suggests reduced hospital LOS and benzodiazepine total dose administered.
MINNESOTA RESEARCH POSTER FINALIST - ALLYSON K PALMER

Targeting Senescent Cells Alleviates Obesity-Induced Metabolic Dysfunction

Authors: Allyson K. Palmer, Yi Zhu, Ming Xu, Tamar Pirtskhalava, Theo H. van Dijk, Esther Verkade, Judith Campisi, Folkert Kuipers, Tamar Tchkonia, James L. Kirkland

Introduction: The prevalence of type 2 diabetes has quadrupled since 1980 and is associated with multi-organ complications including cardiovascular and renal disease. Adipose tissue inflammation and dysfunction are associated with insulin resistance and obesity-related metabolic dysfunction, however mechanisms responsible for this relationship are unclear. Senescent cells accumulate in adipose tissue of obese and diabetic humans and mice, but it is unknown whether they are merely associated with diabetes or if their presence is a causal driver. Cellular senescence is a cell fate that entails proliferative arrest and acquisition of a pro-inflammatory senescence-associated secretory phenotype (SASP). Although senescent cells exist in relatively small numbers in any particular tissue, they have been associated with multiple diseases of aging and are emerging as useful therapeutic targets for age-related diseases, including cardiovascular disease, pulmonary fibrosis, neurodegeneration, and osteoporosis. The SASP of adipose-derived senescent cells includes anti-adipogenic, pro-inflamatory, and chemoattractant factors. We hypothesized that senescent cell removal from diet-induced obese animals could improve metabolic phenotypes.

Methods: In these studies, we induced obesity in mice by feeding a high fat diet, which results in an increased burden of senescent cells in adipose tissue. We then used two methods to eliminate senescent cells. First, we targeted senescent cells in transgenic mice that express a killing gene induced by the promoter of p16^{Ink4a}, a commonly used marker of senescent cells. Secondly, we used drugs that target pro-survival pathways active in senescent cells, termed senolytics. Specifically, we used dasatinib and quercetin (D+Q), which have been shown to cause apoptosis in senescent cells without significant effects in quiescent or proliferating cells. Following senescent cell clearance, we assessed metabolic function by performing glucose tolerance testing and insulin clamping. We also assessed adipose tissue distribution and cellular composition, and tested the effects of senescent cell clearance on macrophage migration into adipose tissue.

Results: We found that genetic targeting in a transgenic mouse model or treatment with senolytic drugs D+Q was able to reduce the burden of senescent cells in adipose tissue of diet-induce obese mice. Senescent cell clearance improved glucose tolerance as well as insulin sensitivity. In addition, adipogenesis was enhanced, adipocyte size was reduced, and adipose tissue distribution shifted toward subcutaneous depots, consistent with improved insulin sensitivity. Macrophage burden and monocyte migration to adipose tissue were decreased after senescent cell removal. In addition, cardiac diastolic function was enhanced and urine microalbuminuria was reduced following senescent cell depletion.

Conclusion: These results indicate that senescent cells play a role in the generation of insulin resistance in diet-induced obesity, and that removal of senescent cells improves the composition and function of adipose tissue, alleviates metabolic dysfunction, and can even impact end-organ complications of obesity. Intermittent dosing of senolytic drugs makes them favorable candidates for clinical translation. Our studies suggest that therapies which target senescent cells represent a novel therapeutic strategy for the treatment of obesity-induced metabolic dysfunction and its complications.
High blood pressure, can we recheck that?

Authors: Andrew Brown, MD, Paul Dotherow, MD, Tim Ryan, MD, Chirag Acharya, MD, Marion Wofford, MD

Introduction: Blood pressure measurement, whether by manual auscultation technique or electronic hybrid devices, is the hallmark of diagnosing and monitoring hypertension. Obtaining accurate blood pressure values is paramount in adjusting medical regimens appropriately. Previously, data indicates inconsistent measurements based on timing, cuff size, patient position, and user training; thus, illustrating the need for a standardized method of blood pressure measurement. Here, we collected two sets of blood pressure data, for out-patient clinic visits at the University of Mississippi Medical Center (UMMC) Internal Medicine Resident Clinic, to test the hypothesis that if patients are allowed sufficient seated time in exam rooms before blood pressure is measured, then measurements may vary.

Methods: For a six-week period from 4/16/2018-5/25/2018, patients seen by the authors during Resident Clinic were included in the study. These patients were checked in under standard clinic protocol with electronic blood pressure measurements upon arrival. Secondly, measurements were obtained as an average of three readings, one minute apart, started at a minimum of five minutes after being seated in the exam room. The overall mean measurements for n=40 patients were calculated and compared using a student’s t-test to determine p-value.

Results: Data from 39 patients was analyzed with means generated and subjected to t-test analysis to generate p-values. The systolic means for control and intervention data points showed an overall difference of -4.82 millimeters of mercury (mmHg); this difference had a p-value of 0.208. The diastolic means showed an average difference of -0.41 mmHg for intervention data points with a p-value of 0.150. Overall, 26 patients had lower systolic pressures after the grace period; 25 patients had lower diastolic pressure. Seven patients had higher systolic and diastolic blood pressures after grace period.

Conclusion: On average, we observed lower systolic and diastolic blood pressures after a grace period of greater than 5 minutes. This difference was larger in systolic (-4.82 mmHg) than the diastolic value (-0.41 mmHg). The difference in neither arm was significant, however, the study needed more power. Regardless, a difference of 5 mmHg in systolic value represents a significant difference warranting further investigation to increase the patient sample size. We also believe this data is a step toward confirming the current literature that the methods of acquiring blood pressure measurements are important for accuracy. Furthermore, the acquisition of blood pressure should be standardized using proper technique, as this affects subsequent treatment decisions.

References

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MISSOURI RESEARCH POSTER FINALIST - ROBIN IMPERIAL, MD

Improving Rate of Screening Mammograms Completed at Truman Medical Center

Authors: Parth Patel MD, Robin Imperial MD, Badar Hassan, MD, Marjan Nazer MD, Waled Bahaj, MD, Ahmed Elkafoaw MD, Julie Banderas, David Wooldridge MD, Sheena Spielberg MD, Kristin Gillenwater, MD

Introduction: Breast cancer is the most common cancer among women in the US irrespective of race or ethnicity. Screening mammography is an essential component to early detection and is recommended by current the US Preventative Services Task Force (USPSTF) guidelines to be conducted biennially for women aged 50-74. Recent statistical analysis shows that 70% of women in the US are compliant with these recommendations. In the Truman Medical Center population, only 45-50% of women are compliant. This study aimed to increase the rate of completed screening mammograms by streamlining the clinic checkout process and education.

Methods: Pre-intervention data (November 20, 2016 to February 18) collected from all 5 internal medicine cohorts. Phase I intervention (Nov 20 – Dec 3, 2017) included front desk education besides providing pamphlets in English and Spanish for every patient with ordered screening mammogram at the time of check out. Those Pamphlets included necessary information about screening mammography available at TMC HH and University health -stating location, office hours, contact number, walk-in option along with a map of office location. Phase II (4-5 Dec 2017) included survey for the patient questioning barriers of not getting the mammogram done. Phase III (Feb 19-March 18, 2018) included medical assistants education. Comparison between pre and post-intervention data was conducted.

Results: In the control group, 50% of patients had the test completed during the pre- intervention period (November 20, 2016 to February 18) compared to 54% of patients during the post-intervention period (November 20, 2017 to March 18, 2018) (p=0.144, 95% CI [-0.109, 0.016]). In the experimental group, 45% of patient had completed the test during the pre- intervention period (November 20, 2016 to February 18) compared to 55 % of patients during the post-intervention period (November 20, 2017 to March 18, 2018) (p=0.001, 95% CI [-0.163, -0.042]).

Conclusion: Compliance rate of mammography is a national problematic issue. Simple interventions like education materials, enhancing the patients’ awareness of its importance and follow up and specific directions to obtain mammograms can significantly increase the rate of completed mammograms. Our study showeda significant improvement in completion following an intervention that could be easily adapted by other institutions.
High-Value Care Education for Third-Year Medical Students

Authors: Paul Kunnath MD, Rachna Rawal MD, Jennifer Schmidt MD, Department of Internal Medicine, Saint Louis University, Saint Louis, MO

Introduction: Medical education literature shows that habits established during training follow physicians through their career. Hence, providing high-value care education to medical students will enable them to recognize and apply high-value care, establishing it as a practice pattern early in their careers. Current state assessment surveys revealed 80% of our institutions incoming third year medical students were not familiar with the term “high-value care”. Additionally, 100% of students identified no prior high-value care education. Our data confirmed what is known in literature—medical students get little exposure to high value care concepts. This presented an opportunity to develop a high-value care curriculum focused at medical students.

Methods: Participants are third year medical students rotating on inpatient Internal Medicine core clerkship. Data on high-value care knowledge is collected by pre and post-intervention survey. Objectives include defining high-value care, distinguishing between low-value and high-value care, and applying evidence-based guidelines to patient care. Interventions includes two educational sessions: an introduction to high-value care during clerkship orientation and a case-based, interactive session mid-way through the clerkship. The material is designed to show students how to consider the risks, benefits and costs of diagnostic testing to make informed diagnostic decisions in an interactive setting.

Results: When students defined high-value care in the pre-survey as a free response, 75% of the free responses included “costs”, “waste”, or “quality” suggesting familiarity with high-value care concepts but lack of more nuanced understanding. Students who felt comfortable utilizing evidence-based medicine for decision making increased from 59% to 91% (statistically significant p<0.05). Post-intervention, 89% of students felt comfortable discussing testing indications with their team, which is a statistically significant increase from 48% pre-intervention (p<0.05). The students identified a lack of knowledge and not having a voice as barriers to engaging in high-value care discussions.

Conclusion: Our data reveals that while students may be familiar with high-value care, they require additional training on its application. After two didactic sessions, students had improved attitudes and confidence in the application of evidence based guidelines. They also showed increase engagement in high-value care discussions with their teams. However, additional initiatives are needed to reduce self-identified hierarchy barriers by empowering medical students and faculty. With early implementation and hands on training in high-value care practices, medical students will establish career practice patterns reflecting high-value care principles.

References

What Happened? Utilizing feedback as a mechanism to improve resident event reporting rates

Authors: Grant A Turner MD, Kristin Lohr MD, Robert B Jones Jr, MD, Emma Lundsmith MD, Megan Margiotta MD, Riti Kanasa-thasan MD, Bracken Babula MD, & Rebecca Jaffe MD

Introduction: Medical errors are responsible for significant patient morbidity and mortality and as a result, increasing voluntary reporting of such events is a priority of the Institute of Medicine (IOM). In line with this, since its founding in 2015 The Housestaff Quality and Safety Committee (HQSLC) at Thomas Jefferson University Hospital (TJUH) has worked to educate housestaff on the value of event reporting. Despite this, event reports entered by housestaff has remained stagnant at ~3%.

Underreporting by housestaff is a common problem in teaching hospitals across the US. Proposed reasons for this include fear of blame and retribution, uncertainty about what should be reported and lack of feedback once an incident has been reported (Jasti et al 2009).

A previous survey of housestaff at TJUH found that 97% of respondents felt the culture for reporting errors was supportive and non-punitive, but only 27% had received feedback on an event they had reported. We hypothesized that improving feedback on event reports would improve rate of housestaff reporting.

Methods: Housestaff event reports at TJUH entered in a 3 month period were confidentially reviewed by the HQSLC. A standardized form was adapted to provide structured feedback to the reporter on timeliness, clarity, objectivity and professionalism of the report. Information on actions taken to address the event were supplied when available. Feedback forms were reviewed by Risk Management prior to distribution back to the reporter via secure email. Recipients were asked to complete a brief survey.

Results: A total of 69 event reports were entered during the pilot period - 38 reporters requested feedback, and this was able to be provided for 30 (79%). Reasons for feedback not being provided included anonymity of the event reporter, follow up information was confidential, or no specific follow up information existed.

Each reporter was asked to complete a brief survey on feedback they received. Quality of feedback was rated on average 3.8 on a Likert scale (1 = not useful, 5 = very useful). 67% of residents felt that the feedback they received encouraged them to report more events in the future, and 50% of residents felt that there was an adequate institutional response to their report. During the studied time period, percent of event reports placed by housestaff increased to 4.5%.

Conclusion: We successfully demonstrated that providing structured feedback on event reports submitted by housestaff helps to promote further housestaff event reporting. While further work needs to be done to streamline this process, it serves as a framework to encourage greater rates of event reporting by housestaff.

References

Cell-free DNA (cfDNA) as a tumor-burden marker in transitional cell cancer (TCC)

Authors: Yen Cao M.D., Mohamed Azab M.D., Subhikshya Tiwari M.D., Mohamad Mubder M.D., Chad L. Cross PhD., Nicholas J. Vogelzang M.D.

Introduction: In current clinical practice, tumor response is mainly determined by radiographic criteria. The assessment of tumor response is challenging as it relies heavily on the nuances of radiographic findings and interpretation. cfDNA has a potential role as a tumor marker represents an inexpensive, noninvasive testing and thus a novel approach to serial sampling for screening and monitoring tumor progression. In this study, cfDNA appears to correlate with tumor burden and thus has potential as a tumor marker in TCC, particularly in regards to determining complete response (CR) after checkpoint inhibitor therapy (CPI).

Methods: This study evaluated patients with solid tumor who undergoing treatment with cFDNA testings. We identified patients with TCC to further assess their cfDNA. The cfDNAs were collected along with corresponding radiographic imaging at the same time interval. We correlated the %cfDNA at each of these time points to the radiographic response of the disease to treatment. Radiographic and clinical responses were categorized into CR, Incomplete Response (IR), Stable Disease (SD), and Progressive Disease (PD). We ran a non-parametric ANOVA based on mean ranks. In patients who achieved CR, we further evaluated disease staging, treatments received duration of CR, and cfDNA percent somatic alteration burden (%SAB).

Results: Of the 197 patients evaluated, .62 had TCC and of these, 24 pts (18:6 M:F, mean age 69) had more than one cfDNA testing (2-7) with a total of 47 tests. 14 ,9 ,8 ,16 of these time points were of pts at CR, IR, SD and PD respectively. Median cfDNA% in CR, IR, SD and PD groups were 0.15, 0.1, 0.8 and 3.65 respectively. There is an overall significant difference when comparing the mean rank of the responses ( p= .005). Bonferroni-corrected multiple comparisons showed a significant pairwise difference between CR and PD (p=.02) and between CR and IR (p=.019). Nine of the twenty-one CPI-treated patients were in clinical CR or near CR and were off CPI for 5+ mos. The %cfDNA for the nine patients were < 0.2 in 7 pts, 0.7 and 0.8 in the remaining 2 patients.

Conclusion: The significant difference in cfDNA% in CR vs PD and CR vs IR suggests an association between cfDNA% and tumor response. More particularly, there is an association between cfDNA%and CR, suggesting a role for cfDNA in disease response to therapy, particularly in CPI. The prospect of cfDNA as a tumor burden marker represents a viable, less-invasive, less-expensive tool to evaluate tumor response and burden when compared to the current radiographic assessment. Further prospective studies with a larger population are needed to evaluate the hypothesis that cfDNA can be used as a tumor burden marker and indicator of tumor response to therapy.
NEVADA RESEARCH POSTER FINALIST - CALEB J MURPHY, MD

Evaluation of novel criteria for identifying wasteful daily lab orders

Authors: Caleb Murphy, Jill Bowman Peterson, Alisa Duran

Introduction: Daily lab testing despite patients’ clinical and lab stability is identified as an area of inappropriate health care spending in the Society of Hospital Medicine’s Choosing Wisely list. Approximately 25% of daily labs are inappropriate, leading to overtreatment and increased spending, yet guidelines for identifying such labs do not exist. The objective of this study was to create and apply criteria to properly identify inappropriate labs compared to the national average of 25%.

Methods: These criteria were created by two academic internists experienced in hospital medicine and high value care, reviewed by the Alliance for Academic Internal Medicine High Value Care Workgroup, and revised. 50 medicine admissions (hospitalization 2-10 days, non-ICU, non-cirrhotic patients) were randomly selected. Using these criteria, two reviewers independently rated appropriateness of basic metabolic panels (BMP) and complete blood counts (CBC) from each hospitalization using both a dichotomous scale (DS; appropriate/inappropriate) and a three-point Likert scale (LS; 1=inappropriate, 2=equivocal, 3=appropriate).

Results: 461 daily labs (253 BMPs, 208 CBCs) from the 50 admissions were reviewed. Using the criteria, 24.1% (95%CI 18.8-29.4%) of BMPs and 25.0% (95%CI 19.1-30.9%) of CBCs were rated inappropriate on the DS. On the LS, 20.2% (95%CI 15.2-25.1%) of BMPs were inappropriate and 7.1% (95% CI 4.0-10.3%) were equivocal, while 16.8% (95%CI 11.7-21.9%) of CBCs were inappropriate and 12.0% (95%CI 7.6-16.4%) were equivocal. When comparing raters on the DS, kappa was 0.68 (95%CI 0.58-0.78) for BMPs and 0.77 (95%CI 0.68-0.87) for CBCs. Weighted kappa on the LS was 0.58 (95%CI 0.49-0.67) for BMPs and 0.62 (95%CI 0.52-0.72) for CBCs.

Conclusion: Using these criteria, raters identified 24.1% of BMPs and 25.0% of CBCs as inappropriate, consistent with previously reported figures. This suggests the criteria correctly identify inappropriate daily lab ordering. When assessed on the LS, inappropriate lab rates dropped as more labs were identified as equivocal. Yet appropriate test rates also dropped; thus, equivocal tests were not just comprised of tests previously identified as inappropriate on the DS. Interrater reliability between raters showed moderate to substantial agreement using both the DS and LS; with more agreement on the DS. These criteria offer an accurate and reliable method of assessing BMP and CBC appropriateness, with potentially important applications in high value care initiatives and medical education. When applied prospectively, this tool could potentially translate to cost savings across health systems and help reduce unnecessary daily lab testing in the hospital.

References

Catheter Ablation versus Medical Therapy for the Treatment of Atrial Fibrillation in Patients with Heart Failure with Reduced Ejection Fraction: An Updated Meta-analysis

Authors: Rajkumar Doshi MD MPH, Krunalkumar Patel MD, Nageshwara Gullapalli MD MPH

Introduction: Current practice guidelines recommend the use of pharmacological rate and/or rhythm control for the treatment of atrial fibrillation (AF) in patients with heart failure with reduced ejection fraction (HFrEF). However, a recent randomized control trial (RCT) has challenged the norm by demonstrating an improvement in survival in these patients when treated with catheter ablation (CA).

Methods: A comprehensive literature search using the SCOPUS database was performed. After examining 410 relevant studies, six RCTs comparing CA with medical therapy (MT) for AF in patients with HFrEF were included. Standard meta-analysis techniques were used to compare all-cause mortality and change in left ventricular ejection fraction (LVEF) between CA and MT. Our primary outcome of interest was all-cause mortality in HFrEF patients with AF, and secondary outcome was change in LVEF. Random effects modeling was used to report risk ratio (RR) and standardized mean difference (SMD) with 95% confidence intervals (CI).

Results: A total of 668 patients among four RCTs were studied for all-cause mortality. The mean age of patients ranged between 55 to 64 years, baseline LVEF from 22% to 43%, and median follow-up duration was between 6 to 60 months. The CA arm had significantly lower all-cause mortality as compared with MT (RR: 0.51; 95% CI: 0.35-0.76, P=0.0009). Restoration of normal sinus rhythm was noted in 69% to 73% of patients in the CA arm as compared with 0% to 34% in the MT arm. Changes in LVEF were reported in six RCTs (N=470). There was an improvement in LVEF in favor of CA (SMD: 0.64; 95% CI: 0.37-0.92, P<0.00001). We observed a significant change in LVEF in patients undergoing CA versus medical rate control (SMD: 0.75; 95% CI: 0.31-1.19, P=0.0008) and a modest change in patients undergoing CA versus medical rate and/or rhythm control (SMD: 0.48; 95% CI: 0.23-0.72, P=0.0001).

Conclusion: CA led to reduction in all-cause mortality and improvement in LVEF in patients with AF and HFrEF. Recent evidence from the CASTLE-AF trial and the results from our updated meta-analysis suggest a paradigm shift is needed for the management of AF with HFrEF. The current practice guidelines may need to be updated to reflect the “strength” of the new evidence and future studies are needed to replicate these findings and to elucidate the pathophysiological basis of the benefits of CA observed in this analysis.
Early Magnetic Resonance Imaging versus Computed Tomography Scan for Evaluation of Cerebrovascular Events in a Community Hospital. - A Cost Analysis

Authors: Geeta Bhagia, M.D.1., Rishi Raj, M.D.2, Amanda Delacruz, M.D.3, Noah Gilson, M.D.4, Stanley Lu, MD5, 1Resident, PGY- 3, Department of Internal Medicine, Monmouth Medical Center, NJ, 2Former Chief Resident, Department of Internal Medicine, Monmouth Medical Center, NJ, 3Resident, PGY- 2, Department of Internal Medicine, Monmouth Medical Center, NJ, 4Chief Department of Neurology, Monmouth Medical Center, NJ, 5Department of Radiology, Monmouth Medical Center, NJ.

Introduction: Stroke is the leading cause of adult disability in the United States and second leading cause of death worldwide. During management of stroke, intracranial hemorrhage is first ruled out by Computed Tomography (CT) scan of head without contrast followed by tissue plasminogen activator administration (if patient qualifies) and/or medical management versus invasive interventions. Recent evidence suggests that a diffusion weighted Magnetic Resonance Imaging (MRI) is equally effective in diagnosing ischemic and hemorrhagic strokes. However, despite strong evidence, many community hospitals continue to perform CT head first followed by an MRI which increases not only the cost and length of hospital stay but also the risk of nosocomial infections and other adverse events. Our study serves to calculate the cost difference (if any) between CT head versus early MRI performed during initial evaluation of stroke.

Methods: We conducted a retrospective chart review of patients who presented to our Emergency department (ED) between 10/01/2015 through 10/01/2017 and required admission for possible cerebrovascular accident (CVA). Inclusion Criteria were Age >/= 18 years, Symptoms suggestive of stroke. Exclusion Criteria were Pregnancy and Age <18 years. We obtained information regarding patients' date of presentation, length of hospital stay, imaging modalities performed and factors prolonging the stay. We calculated average duration of hospital stay, total cost of hospital stay and individual cost for each investigation. A cost analysis was performed. Research protocol was formally reviewed and approved by Institutional review board at Monmouth Medical Center on 12/14/2017.

Results: Study included 8,182 patients. Of those, 8,170 (99.85%) had CT head without contrast and 26 patients (0.31%) got CT with contrast. MRI studies were done in 828 (10%) patients. Total of 634 (7.76%) patients got MRI brain without contrast, 261 (3.2%) had MRI Brain with and without contrast, 406 (4.9%) had MRA head without contrast, 60 patients (0.73%) had MRA neck without contrast, 272 (3.3%) had MRA neck with and without contrast and 1 patient (0.01%) had MRA head with and without contrast. Total days of hospital stay for all patients were 1,797. Average length of stay per patient was 1.9364 days. Calculated health care cost was $25,383,983. Average cost of hospital stay per patient would be $25383983/828= $30656.98. Average per day cost for all patients would be $25383983/1.93days = $13,152,322.8.

Cost Summary: Combined costs of all types of MRI/MRA performed on our 828 patients = $1,413,014. However, if MRI brain with and without contrast was considered as an initial modality for total of 828 patients, the total cost of diagnostic work up would have been = $890,100. Total of $522,914 can be saved on total number of investigations performed and $12,231,660 can be saved by reducing the length of stay to one day for these patients.

Conclusion: We conclude that CT head at the initial evaluation of stroke prolongs the hospital stay (by 1.93 days in our study) at additional health care cost. However, early MRI (in appropriately selected patients) can cut the cost (by $522,914) and length of hospital stay. Further workup can be completed as outpatient if required. However, more studies are required to develop appropriate patient selection criteria.
References


NEW JERSEY RESEARCH POSTER FINALIST - HARSHE MEHTA

Demand Ischemia is Associated with Increased Mortality in Patients with Gastrointestinal Bleeding: Insights from the Nationwide Inpatient Sample 2016

Authors: Harsh Mehta, Ishani Shah, Saad Amin, Abhishek Bhurwal, Akhtar Amin, Mohit Pahuja, Keith Hawthorne

Introduction: Gastrointestinal bleeding (GIB) is associated with a hypovolemic state that can lead to myocardial ischemia. Activation of the sympathetic nervous system as a result of hypotension further increases work of the heart, thereby leading to an imbalance in the demand-supply status of the myocardium, popularly termed as ‘demand ischemia’ (DI). The aim of our study was to provide an analysis of demographic, interventional, comorbid, and patient outcome data in patients admitted with GIB who developed demand ischemia.

Methods: A retrospective cross sectional study of adults (≥18 years) admitted with gastrointestinal bleeding during the year 2016 was performed using the Nationwide Inpatient Sample (NIS) using ICD-10 data. We compared outcomes of patients with GIB (n=7,303,509) developing demand ischemia with patients who did not have demand ischemia. Descriptive analyses were compared using the t-test for continuous data and chi-square test for categorical data. Primary endpoints of our study were in-hospital mortality and length of stay. Statistical significance was assigned at P<0.005.

Results: No significant demographic variations involving age, gender and race were noticed between the two groups. Patients admitted with GIB were more likely to develop demand ischemia when they had additional comorbidities such as heart failure (41.39% vs 20.6%; p<0.005), diabetes mellitus (41.39% 32.6%; p<0.005), chronic kidney disease (41.8% vs 26.2%; p<0.005) and atrial fibrillation (34.84% vs 25.85%; p<0.005). Additionally, patients in the DI cohort were more likely to receive blood transfusion (42.83% vs 26.67%; p<0.005) and were more likely to be critically ill as was evident from use of vasopressors (9.22% vs 4.63%; p<0.005) and development of acute respiratory failure (10.45% vs 3.05%; p<0.005). Overall inpatient mortality was higher in the DI group (3.07% vs 1.16%; p<0.005). On multivariate analysis, having DI was independently associated with increased length of stay in GIB patients (Coefficient 0.12; 95% CI [0.04-0.19]; p<0.005).

Conclusion: Presence of chronic cardiovascular conditions such as heart failure, diabetes mellitus, chronic kidney disease and atrial fibrillation were more common in patients hospitalized with GI bleed who developed demand ischemia. These patients were also more likely to be critically ill and had poorer outcomes as evidenced by higher inpatient mortality and increased length of stay as compared to GI bleed patients who did not develop demand ischemia. Although further studies demonstrating cardiovascular outcomes in this patient population would provide more information, it is imperative that GI bleed patients with demand ischemia are closely monitored during their hospital stay.
Bedside rounds improved patient satisfaction in a community-based internal medicine residency program

Authors: Chinelo Okigbo, Pavan Ganapathiraju, Aileen Hocbo, Snigdha Kanakamedala, Amulya Dakka, Paolo Tempongko, Krizelle Garde

Introduction: The changing nature of hospital practice, with targets to decrease length of stay and increase efficient electronic documentation, has increased administrative time demands on physicians and decreased the time spent at their patient’s bedside. Thus, bedside rounds, one of the important teaching modalities during residency, is on the decline as many residency programs engage in teaching in conference rooms where they can efficiently round, document, and provide teaching on patients on the teaching service. However, evidence suggests that the rise in physician burnout and lack of meaning in the work they do may be linked to the lesser time spent at the patient’s bedside. We postulate that having bedside teaching rounds will increase residents’ connection with their patients and in turn increase patients’ satisfaction with the care received. This study aimed to examine the effect of bedside rounds on patient satisfaction.

Methods: This prospective study used a pre/post intervention study design to obtain data from patients admitted to two teaching services in two separate campuses of a community hospital in southern Jersey (City Campus [CC] and Mainland Campus [MC]). Six patients from each campus were randomly selected per week during the study period to be included in the study (response rate=99%). A total of 60 patients in each campus were studied in the pre-intervention period and 42 patients in each campus were studied in the post-intervention period. Data collected included patient’s demographic factors (age, gender, and preferred language) together with questions assessing patients satisfaction with the care provided by their resident physicians. The intervention was bedside rounding. Descriptive and bivariate data analyses were conducted using Stata statistical software.

Results: The patients’ demographic factors were not statistically different between the pre and post intervention periods. The ability of the patients to identify their resident physicians increased during the study period in both campuses (CC: 35% to 81%; MC: 43% to 83%). Patients who agreed that they received daily updates from their resident physicians also increased in both campuses (CC: 55% to 69%; MC: 65% to 86%). During the pre-intervention period, a greater proportion of the patients perceived that the estimated time spent by their resident physicians at their bedsides was about 10 mins. However, in the post-intervention period, a greater proportion of the patients perceived that the estimated time was 20 or more minutes. The proportion of patients satisfied with the care they received remained the same in the CC (80% to 81%) but increased in the MC (70% to 93%).

Conclusion: This study showed that rounding at the patients’ bedside had positive effects on patients’ knowledge of the daily services received, time spent by their resident physicians at their bedside, and their overall satisfaction with care they received.
Impact of Open versus Closed Intensive Care Unit (ICU) System on Hospital-Acquired Infection

Authors: Ahmad Sharayah MD, Ramy Osman MD, Nasreen Shaikh MD, Noor Hajjaj MD, Sharon Weiner MD, Margaret Eng MD

Introduction: In a “Closed” ICU model, patient is evaluated and admitted under an Intensivist and orders involving patient care are written by the ICU team. While in an “open” ICU model the patient is evaluated and admitted under a primary care physician with Intensivist following the patient as a consultant, orders are written by consultants directly. In a study by ElKerk et al, it was found that a closed ICU system was associated with a 25% reduction in central line-associated bloodstream infection (CLABSI). We sought to identify if any such improvement in CLABSI and other hospital-acquired infections were seen at our hospital since the institution of a closed ICU system.

Methods: Retrospective data analysis on rates of CLABSI, Catheter-Associated Urinary Tract Infection (CAUTI), Methicillin-resistant Staphylococcus Aureus (MRSA) blood infection, Clostridium Difficile (C. Diff) infection and Ventilator-Associated Pneumonia (VAP) was performed in a community medical center under two different ICU models. At our institute ICU was transitioned from “Open” to “Closed” model in June 2016. Infections rates for the period July 2014 to June 2016 when ICU was under the open model were compared to the period from July 2016 to June 2018 when ICU was under the closed model. Data was collected as part of the hospital's infection control surveillance program under standardized hospital surveillance quality monitoring by the infection control team. Dependent T test was used to evaluate statistical significance between the above-mentioned infections under the open and the closed models. P values of (< .05) were considered statistically significant.

Results: There was a 19.3% reduction in CLABSI rate (1.71/1,000 catheter-days in open ICU vs 0.33/1,000 catheter-days in closed ICU, P value: 0.04), 100% reduction in CAUTI rate (2.1/1,000 catheter-days in open ICU vs 0/1,000 catheter-days in closed ICU, P value: 0.03), 100% reduction in VAP (1.9/1,000 ventilator-days in open ICU vs 0/1,000 ventilator-days in closed ICU, P value 0.02). However, there was no significant change in the rate of C Diff infections (1.49/1,000 patient-days in open ICU vs 2.94/1,000 patient-days in closed ICU, P value: 0.87) and MRSA blood infection (0.38/1,000 patient-days in open ICU vs 0.44/1,000 patient-days in closed ICU, P value: 1)

Conclusion: Our study suggests that a closed ICU model is associated with significantly reduced rates of CLABSI, CAUTI, and VAP. It can be speculated that with systematic delivery of care under a single centralized leadership, infectious complications can be significantly reduced.
Mortality Rate of Patients with Septic Shock as Predicted by Number of Vasopressors Required: A Retrospective Observational Study

Authors: Ahmad Sharayah MD, Sinduja Korem MD, Ramy Osman MD, Hanna Sisti PA-S, Noor Hajjaj MD, Chandler Patton MD, Sharon Weiner MD

Introduction: The mortality rate of septic shock is more than 50% in the Intensive Care Unit (ICU), recent studies showed patients with septic shock who received high dose vasopressors (i.e: Norepinephrine ≥0.7 μg/kg per minute) had an increased ICU mortality and decreased survival rate [1]. To our knowledge, there is no study to demonstrate the prognosis of patients with septic shock correlated to the number of vasopressors required to maintain mean arterial blood pressure more than 65mmhg.

Methods: Retrospective data analysis of 131 patients with median age of 68 and Male to female ratio of 61:70 admitted to ICU with septic shock between the periods July 2015 and June 2017 was performed in a community-based medical center. Data was collected using ICD9 and ICD10 codes. Patients’ baseline characteristics including age, gender and comorbidities were collected along with the number of vasopressors used for more than an hour at any point during the ICU stay regardless of the dose. During this period of time ICU was under two models; open and closed model equally. The primary outcome was death at 10, 30 and 90 days from the septic shock onset. Patients who transitioned to hospice care only or patients who did not die due to septic shock were excluded from the mortality analysis. Linear regression analysis was performed with P value of <0.01 being considered statistically significant.

Results: There was significant correlation between the number of vasopressors required and mortality rate in both models of ICU. For patients requiring one vasopressor, mortality rates were 13% in the first 10 days, 18% in the first 30 days and 22% in the first 90 days. For patients requiring two vasopressors, mortality rates were 35% in the first 10 days, 48% in the first 30 days and 50% in the first 90 days. For patients requiring three vasopressors, mortality rates were 57% in the first 10 days, 73% in the first 30 days and 80% in the first 90 days. For patients requiring four vasopressors, mortality rates were 80% in the first 10 days, 100% in the first 30 and 90 days, P value was <0.01 for all of the mortality rates mentioned.

Conclusion: Our study suggests that for patients with septic shock in the ICU, the mortality rate correlates to the number of vasopressors required to maintain mean blood pressure more than 65mmhg, the more the vasopressors needed the higher the mortality rate. The mortality rate for patients requiring four vasopressors is approximately 100% within the first thirty days from the onset of septic shock. Such prognostication is important to the physicians, patients and their families to guide end-of-life care goals.

References


25215094; PubMed Central PMCID: PMC4129888.
Factors Affecting FRAX Score Calculation and Treatment in Practice

Authors: Navneet Kaur MD, Avneet Vig MD, Beverly Johnson MD, Tony Francis MD, Barbara Mendez-Agrusa MD, Jacobi Medical Center, Bronx, NY

Introduction:

- Osteoporosis related fractures cause significant morbidity and mortality. The FRAX algorithm uses clinical risk factors and country-specific fracture data in addition to Bone Mineral Density (BMD) to quantify a patient’s 10 year probability of a hip or major osteoporotic fracture.
- Treatment is recommended for patients with a 10 year risk of ≥ 3% for Hip Fracture or ≥ 20% for Major Osteoporotic Fracture.
- We noticed discrepancies between radiologist reported and physician calculated FRAX score at our hospital. So we hypothesized that all providers are calculating FRAX score differently as BMD in the FRAX calculator is an optional input variable.
- This project was initiated to see the differences in the result when FRAX score is calculated using T-score, BMD and no BMD and how this difference can influence treatment.

Methods:

Retrospective chart review was done of 1200 DEXA reports from 2013 to 2015.

**Inclusion criteria:** Patient between the age of 40-90 years with T-score between -1 to -2.5 at femoral neck.

**Exclusion criteria:** T-score < -2.5 or > -1, patients already on osteoporosis therapy.

Risk factors were obtained from chart review

237 patients met the inclusion criteria.

**Following FRAX scores were calculated.**

<table>
<thead>
<tr>
<th>BMD FRAX</th>
<th>Using femoral neck BMD reported by Hologic DEXA machine (GOLD STANDARD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>T-score FRAX</td>
<td>Using T-score reported by Hologic DEXA machine</td>
</tr>
<tr>
<td>No BMD FRAX</td>
<td>Scoring without using a BMD value.</td>
</tr>
<tr>
<td>Reported FRAX</td>
<td>Scoring reported by the radiologist.</td>
</tr>
</tbody>
</table>

Results: Out of 237 patients, 226 (95.3%) were females. Average age was 67 years. 54.8% were Hispanic, 29.9% Black, 6.3% Asians and 8.5% Caucasians. Following results were obtained using paired t-test and McNemar’s test.
Difference in absolute FRAX score values (Paired t-test)

<table>
<thead>
<tr>
<th>Comparison</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>BMD FRAX vs. T-score FRAX</td>
<td>&lt; 0.0001</td>
</tr>
<tr>
<td>BMD FRAX vs. no BMD FRAX</td>
<td>&lt; 0.0001</td>
</tr>
<tr>
<td>BMD FRAX vs. Reported FRAX</td>
<td>&lt; 0.0001</td>
</tr>
</tbody>
</table>

Treatment difference based on FRAX score (McNemar’s test)

<table>
<thead>
<tr>
<th>Comparison</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>BMD FRAX vs. T-score FRAX</td>
<td>p=perfect agreement</td>
</tr>
<tr>
<td>BMD FRAX vs. no BMD FRAX</td>
<td>&lt; 0.0001</td>
</tr>
<tr>
<td>BMD FRAX vs. Reported FRAX</td>
<td>0.4142</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>FRAX Scores Compared</th>
<th>Discrepancy in treatment</th>
<th>Over treated</th>
<th>Undertreated</th>
</tr>
</thead>
<tbody>
<tr>
<td>No BMD vs. BMD FRAX</td>
<td>38/237 (16%)</td>
<td>38</td>
<td>0</td>
</tr>
<tr>
<td>Reported FRAX vs. BMD FRAX</td>
<td>6/237 (2.4%)</td>
<td>2</td>
<td>4</td>
</tr>
</tbody>
</table>

Conclusion

- FRAX score calculation without BMD leads to both statistically and clinically significant overtreatment especially in elderly.
- Interchanging T score and BMD to calculate FRAX score leads to same treatment decision despite a statistically different absolute FRAX score value.
- Many providers are not aware that if BMD column is left blank, it automatically defaults the calculation to a no BMD FRAX score.
- A pop up alerting the user “no machine was selected so the calculation will default to no BMD which can lead to overtreatment” in the FRAX tool might be helpful to avoid the miscalculation.
NEW YORK RESEARCH POSTER FINALIST - ADITI BHAGAT, MD, MPH

Outcomes of Functional Testing versus Invasive Cardiac Catheterization for the Evaluation of Intermediate Severity Coronary Stenosis Detected on Cardiac Computed Tomography Angiography

Authors: Aditi Angela Bhagat, MD, MPH – Stony Brook University Hospital, Olufunmilayo Agunloye, BS- Stony Brook University Hospital, Getu Teressa, MD, PhD - Stony Brook University Hospital

Introduction: Coronary Computed Tomography Angiography (CCTA) is a non-invasive imaging modality with high sensitivity and negative predictive value for the detection of coronary artery disease (CAD). The main limitations of CCTA are its poor specificity and positive predictive value particularly for lesions of intermediate severity (ICS), as well as its inherent lack of physiologically relevant data on the hemodynamic significance of coronary stenosis. Consequently, acute chest pain patients with ICS receiving a CCTA undergo downstream stress testing or invasive coronary angiography (ICA) to determine the functional significance of the lesion. However, the comparative effectiveness of the two modalities for evaluation of the hemodynamic significance of ICS detected on CCTA is currently unknown.

Methods: We retrospectively reviewed 6,162 CCTAs done in a single academic hospital between the years of 2012-2014. We included acute chest pain patients with a non-ischemic initial electrocardiogram, normal cardiac troponins, and no prior CAD. Of these patients, 118 were identified with ICS (defined as 50-70% stenosis) and either proceeded to an initial stress test (80/118) or an initial catheterization (38/118). The primary outcome was 30-day major adverse cardiac event (MACE) (acute myocardial infarction [AMI], revascularization with Percutaneous Coronary Intervention [PCI] or Coronary Artery Bypass Graft [CABG], and mortality). Secondary outcomes were length of stay (LOS), cardiac catheterization without evidence of significant CAD and therefore no revascularization, and return to hospital for AMI or urgent revascularization.

Results: Among all patients enrolled, females comprised 37%, whites comprised 83%, and the mean age was 57.6 years old. There was no statistically significant difference between those who received an initial stress test in comparison to those who received a catheterization with respect to baseline characteristics including age, race, gender, cardiac risk factors (hypertension, hyperlipidemia, smoking status, family history of premature CAD, diabetes, body mass index). Furthermore, there was no difference in weekend presentation, coronary calcium score, or number vessels involved in ICS. Patients who received a cardiac catheterization had a higher rate of MACE events (44.7% vs. 3.8%, $P <0.0001$) and higher rate of catheterization without revascularization (55.3% vs. 12.5%, $P< 0.0001$) as opposed to those who had an initial stress test. However, there was no difference in hospital readmission for AMI or revascularization and LOS.

Conclusion: Among patients who received a CCTA and were found to have ICS, those referred for an initial cardiac catheterization compared to those referred for a non-invasive stress test had a higher overall rate of MACE and higher rate of negative cardiac catheterization. There was no difference in LOS and 30-day readmission for AMI or urgent revascularization. Therefore, an initial non-invasive strategy may prevent unnecessary revascularization and improve cardiac catheterization yield without negatively impacting LOS and short-term hospital readmission for AMI or urgent revascularization.
Immune reconstitution inflammatory syndrome associated hospitalization in the United States

Authors: Momcilo Durdevic MD, Andreea Constanta Stan MD, Seema Singh MD, Arlene Yu MD, Prakash Acharya MD, Ashutossh Naaraayan MD, Stephen Jesmajian MD

Introduction: Immune reconstitution inflammatory syndrome (IRIS) represents a spectrum of inflammatory disorders, associated with paradoxical worsening of preexisting infectious processes, after initiation of antiretroviral therapy (ART) in HIV-infected individuals. Epidemiological data for IRIS in the United States is lacking. With the addition of specific diagnostic code for IRIS it was made possible to better understand epidemiological specifics of the syndrome. The objective of the study is to describe the epidemiology of IRIS related hospital admissions in the United States.

Methods: We conducted a descriptive, retrospective study on the National Inpatient Sample (NIS) databases for the year 2016. Admissions with HIV and IRIS were selected based on International Classification of Diseases-Tenth Revision, Clinical Modification diagnosis codes (B20 and D893 respectively). Complex survey design, weights, and clustering were accounted for during analysis. Multivariate regression analysis was performed to determine the relationship of mortality and length of hospitalization with IRIS in HIV patients.

Results: The incidence of IRIS related hospitalization is 13.2 per million admissions and the mean age of patients is 43.92 ± 14.67 years. IRIS related admissions were significantly more common in males [OR 4.56, CI (2.83-7.35), p<0.001]. IRIS accounts for 0.25% of HIV related admissions. In HIV patients, mycobacterium avium-intracellulare (16.39%), cytomegalovirus (16.39%), pneumocystis pneumonia (PCP) (13.11%), progressive multifocal leukoencephalopathy (11.48%), cryptococcal infection (9.84%) and Hepatitis B (9.84%) are the most common infectious conditions associated with IRIS. Among HIV admissions with concomitant IRIS, the adjusted odds of mortality is significantly higher [OR 3.14, CI (1.21-8.14), p<0.019] when compared to HIV patients without IRIS. The difference in mortality is mainly due to significantly higher odds of mortality in HIV patients with concurrent PCP infection and IRIS [OR 32.74, CI (4.47-239.76), p<0.001]. The length of hospitalization is 8.49 days longer for HIV patients with IRIS when compared to HIV patients without IRIS [CI (3.8-13.18), p<0.001].

Conclusion: IRIS related hospitalization although rare, is a syndrome that increases the odds of mortality and length of hospitalization in the HIV patients. PCP associated IRIS is the most fatal among HIV patients. The data from IRIS hospital admissions will increase in the upcoming years and will give us the opportunity to assess trends and specifics of the disease.
Safety and efficacy of systemic thrombolytic therapy in pregnancy complicated by pulmonary embolism: an analysis of the nationwide inpatient sample.

Authors: Mohammad I Ghanbar¹, Yuzhou Lou¹ and Adil Shujaat².¹ Department of Internal Medicine, Icahn School of Medicine, Mount Sinai St Luke’s-West Hospital, New York NY., ² Division of Pulmonary and Critical Care Medicine, Icahn School of Medicine, Mount Sinai St Luke's-West Hospital, New York, NY.

Introduction: Pulmonary embolism (PE) during pregnancy is the sixth leading cause of maternal mortality in the US. Whereas the use of systemic thrombolytic therapy (TT) for hemodynamically unstable PE is the standard of care in the non-pregnant population, its use in pregnancy is limited due to concerns of maternal and fetal complications including major bleeding. There is limited literature on its use and no large cohort study.

Methods: The Nationwide Inpatient Sample (NIS) database was used to identify 50,270,033 women with pregnancy-related codes admitted from 2010 to 2016. Pregnant women diagnosed with PE were identified and outcomes were compared between those treated with TT and those not treated with such therapy. Analysis was done using STATA 2015.

Results: 4,352 pregnant women with PE were identified, of whom 71 were treated with TT, which represents a 20% stratified sample size making the real estimation to be around 21,760 for PE cases and 355 for those treated with TT. The mean age of the TT group was 28 years, 56% were Caucasian, and 69% were admitted to a teaching hospital. There was no case of intracranial hemorrhage or antepartum bleeding in the TT group, compared to 0.09% (0.03% - 0.2%) and 0.5% (0.4% – 0.8%), respectively, in the other group. The rate of post-partum bleeding was similar between the two groups at 8% (4% - 17%) compared to 9% (9%-11%). However, the rate of blood transfusions was higher at 23.60% compared to 8.91% (p<0.01). There was no case of abortion in the TT group compared to only 12 cases in the other group. Significantly, more patients in the TT group required intensive care unit (16.7% vs. 5.37%) and intubation (15.28% vs. 5.04% ; p<0.01). There was no significant difference in the use of vasopressors. The mean hospital length of stay was prolonged in the TT group (7.77 vs. 5.41 days; p<0.01). The mean maternal mortality rate was higher in the TT group (12.66% vs.1.37%; p<0.01).

Conclusion: To the best of our knowledge this is the largest cohort of pregnant women with PE treated with systemic thrombolytic therapy. The rates of peri-partum bleeding and fetal demise were not higher in those treated with such therapy; however, the need for blood transfusion was greater with such therapy. Nevertheless, maternal mortality was high despite the use of thrombolytic therapy.
Prognostic significance of JT interval for risk prediction of cardiovascular disease among individuals with normal vs prolong QRS complex

Authors: Reza Mohebi MD\textsuperscript{1}, Aaron Grober\textsuperscript{2} MD, Ayesha Jehan\textsuperscript{3} MBBS, Victor Froelicher\textsuperscript{2,3} MD, \textsuperscript{1}Icahn School of Medicine at Mount Sinai/ NYC Health+Hospitals/Queens, Jamaica, NY, \textsuperscript{2}Center for Inherited Cardiovascular Disease, Stanford University, Stanford, CA, \textsuperscript{3}Cardiology Division, Veterans Affairs Palo Alto Health Care System/ Stanford University, Palo Alto, California.

Objective: Since QRS duration and QT interval have been demonstrated to have independent prognostic value, we hypothesized that separating depolarization (QRS duration) and repolarization (JT interval) into individual components would enhance the prognostic power of these ECG measurements.

Methods: Participants include 16,962 veterans excluding those with no history of myocardial infarction, atrial fibrillation who had an initial ECG at the Palo Alto Veterans Affairs (VA) Health Care System between March 31, 1987, and December 20, 1999, and were followed for cardiovascular (CV) death for 17 years. Linear regression analysis was used to evaluate how QT correction formulas were correlated with its components (QRS and JT interval) and heart rate. Cox proportional hazard regression analysis was performed to estimate the hazard ratio of the corrected QT interval, corrected JT interval and the QRS interval for risk prediction of cardiovascular death.

Results: During median of 17.5 years of follow-up, 481 cardiovascular deaths occurred. Comparison of mean JT intervals among those with normal and abnormal ventricular rhythms showed that the latter group had significantly lower JTc intervals (312 msec vs 298 msec). JTc interval was inversely correlated with QRS interval. The QTc interval was mostly dependent on the JTc interval rather than the QRS segment. However, prolongation of both the JTc and QRS increase the risk of QTc prolongation with similar odds ratios. Among those with normal ventricular conduction rhythms, prolongation of the JTc (HR; 1.12(1.08-1.15)), QRS (HR;1.27(1.21-1.39)) and QTc(HR; 1.13(1.10-1.16)) increased the risk of CVD. Among those with ventricular conduction abnormalities, increases in JTc interval did not increase the risk of CVD (HR; 0.90(0.85-1.14)), however, increase in QRS and QT was associated with increased risk of CVD.

Conclusion: Individuals with ventricular conduction abnormalities had significantly shorter JTc intervals compared with the general population. In that cohort, increases in JTc did not increase the risk of CVD, however, prolongation of QRS and QTc was associated with increased risk of CVD. Among individuals with normal ventricular rhythms, prolongation of all three intervals, the JTc, QRS and QTc, increase the risk of CVD.
An Examination of Medical Malpractice Claims Involving Physician Trainees

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Introduction: Harm events in healthcare are common and place tremendous burden on providers. This phenomenon is called the “second victim effect” and can influence the way providers practice for years after the event. Physicians-in-training may experience substantial emotional burden when involved in harm events during their training. We aimed to determine the factors putting physician trainees at risk of being involved in patient harm events that end in malpractice claims, in order to put in place preventative strategies that target the highest risk areas.

Methods: We designed a case-control study using medical malpractice claims closed between 2012-2016 from the Comparative Benchmarking System. This database is operated by Harvard’s malpractice insurer and contains >30% of the malpractice claims filed in the United States. There is robust quality assurance. The authors identified claims from teaching institutions in which physician trainees were directly involved in the harm events through a coded field called the Service Extender Flag. A control group was formed of claims from the same teaching institutions that did not involve physician trainees. The exposure was a combination of factors, including care setting, primary responsible service, and whether or not a procedure was involved. The main outcome was physician trainees being involved in harm events that result in malpractice claims.

Results: Of 30,973 claims, there were 581 cases with physician trainees involved in the harm event, as denoted by the Service Extender Flag, and 2,610 control claims. Case claims involved residents only (81%), fellows only (13%) or both residents/fellows (6%). Thirty two percent of case claims had trainees named as defendants compared to 9% of control claims (p<0.0001). The most common final diagnosis for trainee involved claims was laceration during surgery (11%). The most common severity of harm for trainee involved claims was permanent injury (36%). Procedures were involved in 71% of trainee involved claims. Inadequate supervision was a contributing factor in the minority of trainee involved claims but was more common in the trainee involved group compared to the non-trainee involved group (24% vs <1%, p<0.0001). Multivariable regression analysis revealed trainees to be at highest risk in procedural fields such as obstetrics/gynecology, especially when performing procedures and delivering care in the emergency room.

Conclusion: Procedural complications and procedural specialties were commonly implicated in trainee involved patient harm events that led to malpractice claims. Inadequate supervision was a contributing factor in only the minority of trainee involved claims. Training directors can use this information to target interventions that will reduce the likelihood of trainees being involved in harm events. This serves the dual purpose of protecting today’s patients from harm as well as tomorrow’s physicians from becoming a "second victim."
Improving PPI prescribing practices in an academic safety-net primary care clinic: quality improvement project

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Introduction: Overuse of Proton Pump Inhibitors (PPI) has increased drastically over the last 2 decades despite evidence based guidelines. The aim of this quality improvement (QI) is to reduce the percentage of patients inappropriately prescribed PPIs between the ages of 40-75 years from a baseline rate of 80% to less than 60% (20% reduction) within 1 year in internal medicine clinic (IMC).

Methods: We used the Plan-Do-Study-Act (PDSA) model and performed a root cause analysis to identify barriers to appropriate use of PPIs. The major barriers included system, provider and patient-based barriers including lack of electronic medical record (EMR) alerts and gaps in knowledge in physician and patients. Multidisciplinary QI team included nursing and ancillary staff, residents, attending physicians and social worker from IMC, technology (IT) department staff and patients.

Outcome measure included reducing rates of patients inappropriately prescribed chronic PPIs. Process measures included the percentage of patients on PPI who have their Gastroesophageal reflux disease (GERD) assessed during the clinic including assessment for alarm symptoms and esophagogastroduodenoscopy (EGD) completion rates in eligible patients. Balancing measures included increase in patient wait times in the clinic and poor access to EGD. QI team performed four PDSA cycles from January 2018 to October 2018. The first PDSA cycle consisted of creation of electronic Health records (EHR) templates with goals of redesigning of nursing workflow to alert physicians to review chronic PPI use and reminding physicians to evaluate alarm symptoms; and to improve medical documentation as a structured data for PPI risk assessment. Other PDSA cycles included education to the physicians, nursing staff and patients. Physician education consisted of PowerPoint Presentation with small group discussion about evidence based guidelines for treatment of GERD. Nursing staff and physicians were also trained on a new EHR template and workflow for chronic PPI assessment. Electronic patient registry was created in collaboration with IT department.

Results: Data analysis was performed by monthly run chart. Patients were on chronic PPI for an average of 3-5 years. Average rates of PPI discontinuation were 30.40% (n=52/171), resulting into 50.0% inappropriate chronic PPI use from the baseline rates of 80% within 10 months. We observed monthly sustainable variations with a median of 28% in discontinuation of PPI. PPI risk assessment was documented as a structured data in 14.6% of patients. EGD completion rates in eligible patients was 46.2% from the baseline of rates of less than 20%.

Conclusion: We exceeded the goal, achieved 30% reduction in inappropriate use of chronic PPI use within 10 months. Lack of automated medical decision support tool was identified as the biggest barrier. Optimization of EHR and education to QI team members were crucial for the success of this QI.
Encounters for Palliative Care, Readmissions and Resource Utilization: Insights from Nationwide Readmission Database

Authors: Karan Sud MD, Sathish Pondaiah MD, Naveen Premnath MD, Vipul Jindal MD, Jennifer Fung MD

Introduction: As health care costs continue to rise, health care systems, and providers are exploring opportunities for cost containment among patients with serious life-threatening illness who incur the highest per capita health care costs. We aimed to assess resource utilization and burden of readmission among patients with encounters for palliative care.

Methods: This is a retrospective study using the National Readmission Database (NRD) for year 2014. All adult patients with an encounter for palliative care were identified using ICD-9 Codes. Patient with mortality in index admission and transfers to other hospitals were excluded to estimate the burden of readmission. Total cost of hospitalization and length of stay were estimated to assess resource utilization.

Results: A total of 157,036 admissions (1.4% of all admissions) were identified with encounter for palliative care for the year 2014. Mean age for the study population was 75 years and 54% were females. Major baseline co-morbidities were Hypertension (60%), Diabetes (25%), chronic lung disease (24%), congestive heart failure (19%), chronic kidney disease (21%), metastatic cancer (12%) and prior stroke (8.3%). Major reasons for index admission among this study group were sepsis (19.4%), acute cerebrovascular disorder (7.0%), congestive heart failure (4.8%) and secondary malignancies (4.8%). In-hospital mortality among this group was 44% and major discharge disposition for remaining patients was skilled nursing facility (48.4%) and home health care (33%). The mean length of stay (SE) and cost of hospitalization (SE) on index admission were 8.8 (± 0.4) days and $23,384 (± 980), respectively. Among patients who survived, 6.7% were readmitted within 30-days of discharge. Major etiologies for 30-day readmission were infectious (17%), neoplasms (15%), circulatory disorders (14%) and respiratory disorders (14%). The mean length of stay and cost of hospitalization on 30-day readmission were 5.9 (± 0.1) days and $12,642 (± 234), respectively.

Conclusion: There is a significant burden of 30-day readmission among patients with encounters for palliative care. Initial longer length of stay and readmissions among this study population contributes to significant resource utilization. More studies are needed to establish the logistical causes of readmission and possible factors that can help contain resource utilization among patients eligible for palliative care.
NEW YORK RESEARCH POSTER FINALIST - AHMED H QAVI, MD

Appropriateness of Troponin Testing in the Emergency Department

Authors: Ahmed H. Qavi MD, Edward Caballero MD, Aireen Kuan MD, Bernard Gitler MD, Stephen Jesmajian MD

Introduction: Troponin levels are routinely ordered within emergency departments (EDs) for the diagnosis of myocardial infarction. It has been suggested that much of this testing is ordered inappropriately leading to overdiagnosis and unnecessary treatment. To our knowledge there has been no published study from the United States regarding appropriate use of troponin in the ED. This study examined the appropriateness of troponin testing within one teaching community hospital in New York.

Methods: A retrospective chart review was conducted of 151 randomly selected patients who received a troponin assay within the hospital's ED. The criteria for appropriateness included chest pain of any nature, breathlessness with pulmonary edema, anginal "equivalent" (breathlessness, nausea, emesis, back pain, palpitations, unexplained hypotension or syncope with new electrocardiogram changes) and new onset atrial fibrillation or flutter. Primary outcome was proportion of patients undergoing troponin testing meeting appropriateness criteria. Secondary outcomes included hospital admissions, acute coronary syndrome (ACS) diagnoses, cardiology consultations and coronary angiographies.

Results: Majority (54%) of troponin testing was appropriate, with the remainder (46%) deemed inappropriate. Of the inappropriately ordered troponins, 20% were positive (range 0.04 to 0.10), and none of those patients were diagnosed with ACS. Only 5% of the total 151 patients were diagnosed and treated for ACS and 2% of them had angiographically proven significant coronary artery disease. Six percent of 151 patients received cardiology consultations. A total of 195 troponin assays were ordered in our 151 patients. Calculating the proportion of inappropriate tests, an estimated total of $128,544 is the projected annual cost of inappropriate troponin testing in our ED.

Conclusion: About half of the troponins ordered in the ED were inappropriate, with no apparent indication to suspect ACS or the need to rule out ACS. These results are comparable to other international studies done in Australia and United Kingdom. Our study will enable physicians to adopt a more rigorous approach when ordering troponin assays. This will result in fewer false positives requiring fewer unnecessary workups for ACS and fewer cardiology consultations for borderline elevations in troponins. These findings necessitate a more robust clinical guideline to develop appropriate indications to order troponins.
Proton Pump Inhibitors Are Associated With Increased Risk of Spontaneous Bacterial Peritonitis in Patients with Cirrhosis: An Adjusted Meta-Analysis of Observational Studies

Authors: Raseen Tariq, Fateeha Furqan, Abdul Wahab, Srishti Saha, Stephen Silver, Sahil Khanna

Introduction: Proton Pump Inhibitors (PPIs) are widely prescribed in patients with cirrhosis for a variety of indications and may be overused. Spontaneous Bacterial Peritonitis (SBP) is a common but serious complication in patients with cirrhosis. Studies evaluating the risk of SBP in patients on PPIs have shown conflicting results. We performed a systematic review and meta-analysis to study the association between gastric acid suppression medications and the risk of SBP.

Methods: A systematic search of Medline, Embase, and Web of Science was performed up to June 2018. Studies (case series, case-control, cohort studies and clinical trials) assessing the association between PPI exposure and SBP in patients with cirrhosis were included. Summary Odds Ratio estimates with 95% confidence intervals (CIs) were calculated with the random-effects model.

Results: Twenty-two studies with a total of 12,265 patients with cirrhosis were included. Of those, 4,748 were exposed to PPIs. The rate of SBP in patients on PPIs was 16.9% (807/4,748), compared to 15.2% (1,147/7,517) in patients not on PPIs. Meta-analysis showed an increased risk of SBP in patients using PPIs with an OR (2.05, 95% CI 1.64-2.56, p<0.0001). There was moderate heterogeneity among the studies with an $I^2$ of 60%. Of the included studies, 19 studies had adjusted for potential confounders. Analysis of studies that had adjusted for potential confounders also revealed increased risk of SBP with the use of PPIs (OR 1.74, 95% CI 1.41-2.16, $I^2$ = 53%). Risk remained significantly high in subgroup analysis of only cohort studies (OR 1.49, 95% CI 1.26-1.76, $I^2$ = 3%).

Conclusion: Meta-analyses of existing studies suggest that use of PPIs is associated with an increased risk of SBP. The risk remains high even after adjusting for potential confounders. It may be reasonable to re-evaluate the need of PPIs in patients with cirrhosis and stop them whenever possible.
NEW YORK RESEARCH POSTER FINALIST - THINZAR WAI, MD

Treatment Outcomes between HCV Mono-infected and HCV/HIV Co-infected patients

Authors: Thinzar Wai; MD, Anca Giurgiulescu; MPH, Belisario Bejarano; MD, Zeyar Thet; MD, Jilan Shah; MD, Jyoti Gupta; MD, Fils-Aime, Stephany

Introduction: CDC data from the national health survey has shown that reported cases of acute HCV infection have been increasing about 3.5 times from 2010 – 2016 period. The primary care-based HCV treatment and outcomes have been one of the best methods in clinical settings and could provide more advantages for HCV patients who also require the extensive management of coexisting chronic infection. This study will evaluate the outcomes of sustained virologic response (SVR) of the Hepatitis C infected patients alone or along with HIV co-infection. SVR rate will be a standard of 12 weeks period after the HCV treatment.

Methods: This is a retrospective study and the data of 101 patients of HCV mono-infected or coinfected with HIV were collected from the electronic medical record from the period of January 1, 2014 to December 31, 2017 at GI and ID outpatient clinics at WHMC. The association between treatment outcomes of sustained virologic response (SVR) among HCV mono-infected and HCV/HIV co-infected patients were evaluated. Descriptive statistics and chi-square were used to analyze the data.

Results: Out of 101 patients, 80% (N=81) were baby boomer groups who were born between the period of 1945-1965, 20% (N=20) were non-baby boomer group. The average age of the patients was 59. Among them, the majority are male 69%, where female was 31%. Similarly, 46% were Hispanic and 38% were non-Hispanic, 17% were refused to answer. Overall, 44% of SVR attained over the four-year period though SVR rate has been declining during the four years’ time frame. The percent of missing data or lost to follow up among only HCV mono-infected patients (27%) was higher compared to those with HCV/HIV co-infected patients (7%). After exclusion of missing data, interestingly, 56% of the HCV/HIV co-infected patients achieved SVR (>12 weeks) whereas 33% of HCV mono-infected patients had attained SVR. Moreover, the p-value was 0.029 which is less than .05, thus we can conclude that there is a significant relationship between HCV mono-infected/HIV co-infected patients with SVR. However, no statistical association was found between genotype and SVR rate (p-value:0.27).

Conclusion: This finding of a significant association between HCV/HIV coinfection and SVR rate will help to monitor SVR rate especially to promote follow-up visits after 12 weeks of the treatment course, along with HIV primary care services. Since there were higher lost to follow-up for HCV mono-infected compared to HCV/HIV co-infected patients, we can conclude that HIV patients were significantly established with their primary care at ID clinics, and more likely to adhere follow-up visits where HCV mono-infected patients will necessitate further approach to create a regular follow-up at least 3 months/6 months/annual visits after the HCV treatment course. This study will help to participate in HCV quality improvement across the state or national level, in addressing a global concern of HCV epidemics.
VTE Prophylaxis in Hospitalized Medicine Patients: Too Much of a Good Thing?

Authors: Soombal Zahid DO, Allison Schure MD, Carissa Windish MD, Rebecca Mazurkiewicz MD, Linda Kirschenbaum MD

Introduction: Prevention of venous thromboembolism (VTE) is a national issue of quality care. Pharmacologic VTE prophylaxis is recommended for intermediate and high risk patients, but not for low risk patients. Several models for VTE risk stratification have been developed, with IMPROVE (International Medical Prevention Registry on VTE) being validated in a multicenter study for use in hospitalized populations. We decided to analyze such risk stratification and pharmacologic prophylaxis use at our hospital.

Methods: We obtained a list of medical floor inpatients from 11/1/2016-01/20/2017, from which risk scores were calculated using IMPROVE. We assessed for prophylaxis decisions and calculated IMPROVE scores using admission documentation. The score was then used to determine if prophylaxis was appropriately prescribed for patients. Overuse was defined as pharmacologic prophylaxis for low-risk patients (i.e IMPROVE score of 0-1). In order to identify barriers for appropriate prophylaxis and gain insight into existing faculty and housestaff knowledge, we administered a 4 question survey to internal medicine residents of all training levels as well as hospitalists. We then created a 4 minute video didactic session discussing proper VTE risk assessment and prophylaxis, referencing ACP and CHEST guidelines. This intervention was followed by a 5 question survey to determine participant rating of session and understanding of concepts.

Results: Out of 146 patient charts, 108 met inclusion criteria. 89% of patients had pharmacologic prophylaxis ordered with a score of zero, and 100% with a score of 1. In terms of survey data, 67 out of 96 respondents (70%) completed the pre-intervention survey. 26 respondents (39%) felt the decision to prescribe prophylaxis was influenced by senior residents or attendings instead of evidence-based risk assessment models; 10 respondents (15%) admitted that only clinical judgment is used to decide; and belief in routine prophylaxis for all hospitalized patients was given as an answer by 3 respondents (4%). The post-intervention survey had 71 respondents (74%), 63 (94.6%) agreed that the educational session was effective in improving comfort, confidence, and understanding of VTE risk assessment. Moreover, 98% of respondents expressed the intention to use a risk model to guide decision making for pharmacologic prophylaxis of hospitalized patients in the future.

Conclusion: Indiscriminate use of pharmacologic VTE prophylaxis for medicine inpatients at low risk for DVT or PE was common at our institution due to a knowledge deficit about the tools available for assessing VTE risk. By educating residents and attendings about a model such as IMPROVE, and reinforcing guidelines on the utility of pharmacologic prophylaxis, we were able to promote high-value care. Post-intervention analysis showed that the didactic session had a positive impact on hospitalist and resident knowledge of appropriate VTE prophylaxis use and also helped with comfort and confidence in using risk assessment models in the hospital setting.
Effects of Medicaid Expansion on Avoidable Mortality in Kentucky in Comparison to North Carolina

Authors: M. Leila Famouri, MD MPH; Julia Rushing MS; Ramon Velez, MD MS

Introduction: In 2014, the Affordable Care Act (ACA) expanded Medicaid eligibility for most adults with incomes up to 138 percent of the poverty level, which resulted in large decreases in uninsured rates in both expansion and non-expansion states, especially in low-income populations. Studies have indicated that Medicaid expansions prior to the ACA have led to decreases in all-cause mortality; however, the ACA Medicaid expansions’ effect on mortality, particularly health care amenable mortality, is less clear. To explore this relationship, we compared health care amenable mortality rates in Kentucky, an expansion state, with North Carolina, a non-expansion state.

Methods: All mortality data were obtained from the CDC WONDER online database, and all demographic data were extracted from the American Community Survey. We defined 2010-2013 as the pre-reform period and 2014-2016 as the post-reform period. Individuals who were not between the ages of 15-64 during the pre or post-reform period were excluded from the analysis. Demographic characteristics and health care amenable age-adjusted mortality rates were collected at the county level. A generalized linear model was developed for age-adjusted health care amenable mortality in both states, adjusting for demographics including median income, gender, marital status, race, and unemployment rate. The model was also adjusted for percent of population on income assistance, on SNAP, below the poverty level, and age above 65. The average age-adjusted mortality rate for the pre-intervention period (2010-2013) was also used as a covariate in the model to ensure that the comparisons were balanced beginning with the intervention period.

Results: All demographic characteristics significantly varied between the two states (p<0.05). Between 2010-2016, age-adjusted health care amenable mortality increased by 6.25% in Kentucky, while North Carolina saw a modest decrease of 0.7% during the same time period. Crude age-adjusted mortality rate in the post-reform period in Kentucky was 184.9 per 100,000 as compared to 176.2 in the pre-reform period. Crude age-adjusted mortality in North Carolina was 148.1 per 100,000 in the post-reform period and 147.6 in the pre-reform period. In the generalized linear model, both states showed increasing age-adjusted mortality rates with time, but Kentucky had a sharper increase (188.92 to 204.76, p-value<0.0001) in comparison to North Carolina (178.61 to 181.84, p-value<0.0001) during the post-reform period.

Conclusion: This analysis suggests that the ACA Medicaid expansions either had no effect or resulted in increased mortality in the state of Kentucky. Given that age-adjusted health care amenable mortality has been steadily increasing in Kentucky since 2010, it is possible that increased insurance coverage has not provided the long-term medical management necessary to amend outcomes in mortality. We plan to conduct additional studies to better characterize the relationship between Medicaid expansion and mortality in these states.
Caregiver Access to Online Patient Portals: Results from a National Survey

Authors: Rachel Wilson, Celine Latulipe, Syeda Fatema Mazumder, Jennifer Talton, Sarah Quandt, Alain Bertoni, Thomas Arcury, Kathryn Melius, David P Miller

Introduction: Patient portals are secure websites that let patients access their personal health information. To facilitate care coordination, some health systems let caregivers create "proxy accounts" to access information of those they assist. For example, an adult daughter might create a proxy account to review her elderly mother's medications and upcoming appointments. However, proxy accounts could jeopardize patient confidentiality if they fail to limit the types of information that can be seen. Additionally, failing to offer proxy accounts could encourage password sharing which can threaten confidentiality. It is currently unknown how many hospitals offer proxy accounts or whether how often limits can be placed on information available to proxies.

Methods: We randomly selected two general medical hospitals from each state and the District of Columbia using the FY2016 American Hospital Association Annual Survey Database™ to create a national sample. A research assistant, posing as the daughter of an elderly woman who was planning to move to the area, called each hospital to ask if the system had a patient portal that could help her manage her mother's healthcare. Our primary outcome of interest was the proportion of hospitals that offered proxy account access for adult patients. Secondary outcomes included whether proxy access could be limited to certain types of information (such as appointments only) and whether the "daughter" was advised to use her mother's password in lieu of creating a proxy account. The study protocol, including the use of deception, was approved by the Wake Forest School of Medicine IRB.

Results: Of 108 contacted hospitals, 94% (102) had a patient portal and were included in the study sample. Of the 102 hospitals, 69 (68%) allowed caregivers to create their own proxy accounts. Independent hospitals were less likely to offer proxy accounts than hospitals within a health system (55% vs. 80%, p=0.01). Only 19% (13/68) of hospitals with proxy accounts allowed patients to limit the types of information available to their proxies. When setting up proxy accounts, 30% (21/69) of hospitals required both the patient and person requesting proxy access to be present together. 45% of hospitals (42/94) actually suggested the daughter log-in as her mother.

Conclusion: Two-thirds of the participating hospitals offer proxy account access. However, few hospitals allow patients to limit the information their proxies can see and many recommend patients share passwords. While patient portals have potential to assist care coordination, as currently implemented, they represent a threat to patient data security. To protect confidentiality, oversight organizations such as The Joint Commission and the Centers for Medicare & Medicaid Services should mandate patient portal protections for vulnerable populations.
The Utility of Liquid-Based Biopsy Using Next Generation Sequencing Technology at the Time of Tissue Collection by Bronchoscopy for Suspected Lung Cancer

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Introduction: The identification of and targeted therapy for actionable mutations in non-small cell lung cancer has improved mortality and quality of life. Traditionally, these genetic alterations are tested in tissue that is often collected by bronchoscopic methods. Frequently, there is insufficient tissue to perform these studies, which may result in delays in therapy and additional invasive procedures. Circulogene™ (Birmingham, AL) is a blood-based molecular test that can detect genetic alterations with next-generation sequencing technology. We aimed to evaluate the utility of Circulogene™ (Birmingham, AL) at the time of bronchoscopy in patients with suspected lung cancer.

Methods: We retrospectively evaluated consecutive patients that underwent a diagnostic bronchoscopy for suspected lung cancer over 12 weeks. Blood was collected at the time of the bronchoscopy and held until the final diagnosis of cancer was rendered. Once a diagnosis of non-small cell lung cancer was confirmed, the blood and tissue samples were sent for next-generation sequencing to identify any actionable mutations. The time from the tissue collection to treatment and concordance of actionable mutations between the blood and tissue samples were compared.

Results: Fifty-six patients were diagnosed with non-small cell lung cancer. Twenty-four patients (43%) had adenocarcinoma, 21 (38%) with squamous cell carcinoma, 10 (17%) with NSCLC NOS, and 1 (2%) with a mixed small cell/squamous cell lung pathology. Blood and tissue samples were evaluated for actionable mutations and 12 patients (21%) expressed a mutation. The blood results correlated 85% (64% to 100%) of the time on average with the tissue samples for an actionable mutation. The average turnaround times for Circulogene™ (Birmingham, AL) results was 7 days, compared to 14 days for the tissue samples. Chemotherapy was initiated on average 17 days after the bronchoscopy.

Conclusion: Circulogene™ (Birmingham, AL) seems to provide accurate identification of actionable mutations in a reasonable time period compared to tissue based testing. We present the first report evaluating the utility of a blood test that employs next-generation sequencing to identify genetic mutations in non-small cell lung cancer collected at the time of bronchoscopy. The impact of liquid biopsy on time to treatment and the need for additional invasive testing needs further evaluation.
Comprehensive look-up table of KRAS through saturation mutagenesis and pooled transformation assay

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Introduction: KRAS is the most commonly mutated proto-oncogene in human cancers. Even though the majority of KRAS mutations occur at hot spot codons such as G12, G13, Q61, and K146, hundreds of additional mutations were discovered in recent years as more tumors are sequenced. These unstudied missense mutations, termed ‘variants of unknown significant’, pose a great challenge in interpreting clinical sequencing data. For example, it is currently a standard of care to use KRAS mutation status as an exclusion criterion for EGFR inhibition therapy in metastatic colorectal cancer. We wanted to construct a comprehensive lookup table for KRAS missense mutants and investigate principles underlying observed frequency of KRAS mutations in cancers.

Methods: KRAS4B cDNA was cloned into pUC57 vector. Saturation mutagenesis was performed with primers incorporating 19 amino acid substitutions in each of 187 codons (excluding start codon). Out of possible 3,553 mutants, more than 99% of the clones were detected in plasmid pools. The plasmid pool was cloned into a lentiviral vector (pLX307). Pooled lentivirus was transduced into immortalized human epithelial cells (HA1E). Genomic DNA was harvested on Day 0 and Day 7 from transduced cells cultured in either high or low attachment plates. Nextera sequencing was performed for deconvolution. Three biological replicates were included. Mean of Day 7 enrichment compared to Day 0 was used as a surrogate for transforming potential.

Results: All previously known transforming alleles of KRAS scored high in our assay. We additionally discovered rare transforming alleles such as Q22F/I, L23G/N/R, D92K/R, and N116L/M/V. Interestingly, there were alleles that promoted growth in high attachment plates but failed to induce transformation, such as mutant alleles of N85, N86, and T87. Among the transforming alleles, the incidence of mutations found in cancers was mostly explained by the strength of transforming potential of the mutation, and the likelihood of that particular mutation to occur. For example, G13P was more transforming than G12 or G12D, but proline requires more nucleotide substitution, resulting in G13P being rarer than the latter two.

Conclusion: We have successfully performed saturation mutagenesis of KRAS and constructed a gain-of-function dictionary of alleles. This resource may facilitate understanding of clinically important mutations. The potency of transforming potential and the likelihood of substitution were the driving force in determining the incidence of KRAS mutations observed in human cancers.
Introduction: Food insecurity, defined as a lack of consistent access to any type of food for all members of the household (1). Proven associations with obesity (1), depression (2), hyperlipidemia (3), diabetes (4), and hypertension (5) have been repeatedly demonstrated. The result is an overall reduction in wellbeing (2), as well as a drastic reduction in life expectancy by, even when controlled for socioeconomic status (6). Food insecurity during childhood is also correlated to food insecurity as an adult (7). Our clinic has a much higher than average prevalence of obesity (75%), diabetes (33%), and hypertension (57%). We enrolled patients with USDA-qualified food insecurity AND diabetes, obesity, hypertension, or hyperlipidemia in a study combining “Prescriptions for Produce” and a Culinary Medicine cooking course.

Methods: We provided a weekly educational class taught by a certified culinary medicine expert (provided by Local Matters) in the Mount Carmel Healthy Living Center Teaching Kitchen. A grant was obtained for $900 from Columbus Public Health Department to provide “Prescriptions for Produce”. This funding subsidized Franklinton Gardens providing $10 of weekly free in-season vegetables, delivered to the location of the course. Pre/post change in total body weight/BMI, PHQ-9 scores, routine lipid panel, systolic and diastolic blood pressure, HgbA1c, Mediterranean Diet Survey, food preparation methods survey (externally validated), and confidence in cooking survey (externally validated) were measured.

Results: Non-statistically significant improvements in weight, depression, cholesterol, and diastolic blood pressure were observed within the 6 weeks of the class. Statistically significant improvements in waist size (p-value = 0.0156), Mediterranean diet scores (p= 0.0156), food preparation methods (p= 0.0455), and confidence in cooking (p=0.0313) were observed.

Conclusion: Our assessments showed significant effects in behavioral change, indicating effectiveness of the educational process, the first step towards sustainable change. Biomarkers have not significantly changed during the 6 week course. A 2015 meta-analysis of studies investigating 56 published papers utilizing lifestyle interventions for obesity had a follow-up sampling range of 4 to 18 months, indicating our 6 week biomarkers are a very early sample (8). Although many biomarkers were not statistically significant after a 6 week intervention, the authors expected interventions to take at least 3 months to alter biochemistry. Future directions may investigate the effect on mental and physical health by holding a weekly family-style, sit-down dinner. The completion of the PRODUCE trial is expected in the next two years. 3 to 6 month follow-up samples are currently being collected. A rolling schedule of classes has been established to facilitate recruitment from the clinic and increase sample size.

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3. Household Food Insecurity Is Associated with Adult Health Status
Impact of a Standardized Note Template in the IMC on Note Completion Time

Authors: Michael Nguyen MD, Kristin Hom DO MPH, Sideris Facaros MD, Michael Oravec MPH, Emily George MD, Rose Penix, MPH, Michael Rich MD

Introduction: Timely provider EMR note completion is important to patient care, clinic efficiency, resident satisfaction, and steady reimbursement. The established standard for the Summa Health Medical Group (SHMG) is that notes should be completed within 24 hours of the encounter. Administrative data noted considerable room for improvement in the Summa Internal Medicine Center (IMC) note completion time, and resident feedback supported the use of a standardized note template. The use of template-guided notes has been associated with improved billing data, decreased clinic visit time, and increased provider satisfaction. The purpose of this QI initiative is to evaluate the impact of a standardized note template that has been implemented in the IMC to improve note completion time.

Methods: The IMC is a residency-based primary care clinic that serves a diverse population of patients with multiple complex medical issues. In July 2017, a standardized note template was introduced to assist residents with their assessment and plan, and improve overall efficiency. Use of the note was optional to residents. To evaluate the impact of this note, we reviewed all completed IMC visits from July 2017 through January 2018. We compared mean time for note completion and the proportion of notes completed within 24 hours for visits that used and did not use the note template. Independent samples t-tests and chi-squared tests were used to conduct bivariate analyses, and multivariate regression models were used to assess difference in note completion time and 24 hour completion, controlling for provider status (resident/faculty/advanced practice provider), resident experience (postgraduate year), and provider workload (daily completed visits).

Results: We assessed 8019 IMC visits, of which 1763 (22.0%) used the template. Mean note completion time was 9.0 hours using the template, compared to 21.8 hours without the template (p<.001). In the template group, 93.9% of notes were completed within 24 hours, compared to 81.9% without the template (p<.001). Adjusting for provider status, experience, and workload, use of the note template was associated with a 10.6 hour reduction in note completion time (p<.001), and an increased likelihood of completion within 24 hours (OR 3.08, p<.001).

Conclusion: Use of the note template was associated with a significant improvement in mean note completion time and 24 hour completion. As only 22.0% of IMC visits have used the template since its introduction, use of the template should continue to be expanded.
**OHIO RESEARCH POSTER FINALIST - JESSICA E WEISS, DO**

**Reduction of Daily CBC/BMP Labs Among Internal Medicine Residents at Mount Carmel West**

**Authors**

Jessica Weiss, D.O., Noah Hagen D.O., Carter Battista D.O., David Aufdencamp M.B.A., Lynn Shafer, Ph.D., Robert Battisti M.D.

**Introduction**

As part of the ABIM Choosing Wisely initiative, the Society of Hospital Medicine has emphasized repetitive complete blood count (CBC) and basic metabolic panel (BMP) testing among its top five practices that physicians and patients should question. Reduction of unnecessary laboratory testing is important not only for cost containment, but also for improved patient experience and avoidance of iatrogenic harm. It was found at Mount Carmel West, the direct charge of a CBC was $36 and BMP was $40. A literature review showed that a quality improvement project utilizing educational interventions including cost education and appropriate utilization of lab testing could be successful.

**Methods**

Our internal medicine residency program is seeking to reduce unnecessary ordering of daily CBCs and BMPs on the general medicine floors with a goal of decreasing CBC/patient/day and BMP/patient/day each by 20%. Interventions consisted of education and feedback and were tailored for internal medicine residents. Education included lectures, monthly reminders, email updates and lab cost badge displays. Residents were encouraged to evaluate each patient daily for the need for a CBC and BMP with feedback regarding each team’s progress provided monthly. This project used pre-intervention data from 07/2015 to 08/2016 to compare with post-intervention data from 09/2016 to 12/2017 in number of CBC and BMP orders per patient/day, which were then compared using the two-sample Poisson rate test for statistical significance.

**Results**

This project was targeting a 20% reduction of CBC/patient/day and BMP/patient/day on the general medicine teams. With our educational intervention, we reduced BMP/patient/day by 23.1% and CBC/patient/day by 10.9%. Even by partly reaching our goal of 20% reduction, we decreased healthcare costs by several hundred thousand dollars per year.

**Conclusion**

Since initial intervention, there has been a total reduction of $282,667 in cost of care. This quality improvement project took only 16 to 20 person-hours and a small cost to make lab cost badge displays. With more time and further education, we believe our goal reduction in CBC ordering can be reached.

**References**

5. Mount Carmel West Charge Database

**pg. 141**
Newly adopted cardiac telemetry monitoring guidelines impact patient care at a large tertiary care center

Authors: LOGAN DALAL MD, JASON SCHOTT DO, ANDREW MACMILLAN DO, BRENDAN KAPPUS DO, ANAS MANSOUR MD, RANI SHAH DO, JOHN O. ELLIOTT PHD MPH, KIM JORDAN MD FACP

Introduction: The American Heart Association (AHA) has provided specific recommendations for the use of cardiac telemetry monitoring. However, telemetry overuse still occurs in the low risk population and with reportedly 20-40% of patients on telemetry monitored without appropriate indication. Overuse may result in excess expense, mismanagement, alarm fatigue, and prolonged hospital stay. Institution-specific guidelines were established, conveyed through educational sessions, and implemented into electronic medical record (EMR) order sets with the aim to improve appropriate use of cardiac telemetry.

Methods: The study was implemented over multiple cycles from 2014 to 2016. The initial cycle consisted of an education focused intervention. Hospitalists, resident physicians, and internal medicine faculty were surveyed on telemetry use practice patterns and asked to provide best response to management of 6 clinical scenarios. A brief educational session highlighting institutional guideline recommendations on telemetry use was provided and participants were re-tested with identical clinical scenarios, on knowledge of appropriate use. The next cycle included the distribution of pocket cards displaying the guidelines, and a refresher presentation to incoming residents. Chart reviews were performed on 200 randomly selected patients who received telemetry prior to and over 6 months after the educational session and pocket card distribution. In the final cycle, institution-specific guidelines for cardiac telemetry initiation were included in admission order sets. Following this intervention, data on telemetry use was again collected on 200 randomly selected patients. Data collected included dates and indications for telemetry initiation, appropriate discontinuation based on institutional recommendations, and adverse events noted.

Results: After the initial educational session, there was a statistically significant increase in appropriately stopping telemetry when no longer indicated, as demonstrated on a case-based survey. In patients with no indication for telemetry use, the number of patients on telemetry decreased from 16.3% to 1.4% following EMR order-set implementation. Mean excess days on telemetry were reduced by 43.4%.

Conclusion: Overuse of telemetry remains a persistent issue. Our project demonstrated that modification of order-sets was most useful in decreasing inappropriate use of telemetry in patients who do not meet AHA criteria for monitoring. This intervention proved to have a benefit on overall telemetry duration, which can be modeled to cost savings. While no specific cost data was available at our institution, at other hospitals within the system, there was a 341-dollar difference per day between telemetry and non-telemetry beds. The study’s limitations included smaller sample sizes, as data collection via manual chart review is time consuming, subject to incomplete data due to poor documentation, and restrains change cycles. Moving forward, we hope to assist with appropriate discontinuation of telemetry through best-practice advisory alerts.

References


Medication reconciliation errors among skilled nursing facilities discharged patients

Authors: Kim H, Monachese M, Kim L

Introduction: ‘High quality transitional care’ is important not only to prevent harm (1) to patients but also to reduce unnecessary medical expense (2, 3, 4). Previous studies have found medication reconciliation errors play a significant role in impacting ‘High quality transitional care’ (5, 6, 7, 8). This study investigated the prevalence of medication reconciliation errors among discharged patients to Skilled Nursing Facilities (SNFs) and whether these medication reconciliation errors impacted 30 day readmission.

Methods: We reviewed the medical records of Cleveland Clinic Main Campus patients who were discharged to SNFs within a 25-mile radius of the hospital for the months of July and December, 2014. Three physicians independently reviewed the medication list of discharge summary and discharge instructions and compared the list of medications in the medicine administration record and prior hospitalization medication list.

Medication reconciliation error was defined as discrepancies including omissions, duplications, dosing errors, or drug interactions which leads to, or has the potential to lead to, harm to the patient (9,10). We defined ‘severe medication reconciliation error’ as among those errors prompting life threatening events and confirmed by 3 physicians’ agreement.

Cleveland Clinic’s Institutional Review Board approved the study and waiver of informed consent.

Results: Overall 218 patients were included in final analysis 98 and120 from July and December, 2014 data respectively. For July, 32(32.7%), patients were discharged with any medication reconciliation errors 12 (12.2%) of which were found to be severe. 27(27.6%) patients were readmitted within 30 days. Medication reconciliation error group had a higher but statistically non-significant 30-day readmissions (34.4% vs 24.2%, P=0.43). For December, 37(30.8%) were discharged with medication reconciliation errors, 5 (4.2%) of which were found to be severe. 23 (19.2%) of patients were readmitted within 30 days. Medication reconciliation error group had a high 30-day readmissions but statistically not significant. (21.6% vs 18.1%, P=0.71).

Conclusion: 69 patients(31.7%) were discharged with medication reconciliation errors in July and December 2014. The medication reconciliation error group had higher 30-day readmission rate, but statistically not significant (27.5% vs20.8%, P=0.39). Further research is needed to investigate whether medication reconciliation error directly affect readmission rates or if medication reconciliation error is a marker for a poor outcome since it reflects patients’ complexity such as multiple transitions or longer hospital stay. Definition of medication reconciliation error is a relatively vague concept as discharge medication list would reflect physicians’ undocumented clinical intention.

References

Reducing Inappropriate Telemetry on the Inpatient General Medicine Teaching Services

Authors: Fatima Shahid MD, Nabil Madhun MD, Shailee Shah MD, Susan Vehar MD, Anne Harwood MD, Rabel Misbah MD, Jason Wheeler MD, Tammara Sussman MD, Lauren Buehler MD, Shruti Gandhy MD, Dianna Copley APRN, Jessica Donato MD

Introduction: Approximately $250,000 is spent on unnecessary cardiac monitoring in an average hospital annually. In 2004, the American Heart Association (AHA) published guidelines for cardiac monitoring; overuse of telemetry results in waste, over-treatment, patient discomfort and alarm fatigue. This resident-led multi-disciplinary quality improvement project was aimed at reducing inappropriate ordering of telemetry by residents rotating through General Internal Medicine (GIM) teaching service by 20% over 3 months by implementing a multipronged intervention targeting resident knowledge of telemetry guidelines and awareness of telemetry status.

Methods: A baseline survey was conducted to assess trainee knowledge regarding indications for telemetry and the associated cost. Review of the electronic medical record was used to determine telemetry status and indications for patients on GIM teaching service between January 2018 and March 2018. Cleveland Clinic Health System Telemetry Guidelines, which are based largely on the AHA Telemetry Guideline’s class I and II indications, were used to determine appropriateness of telemetry orders. After baseline data collection, a series of interventions were implemented. In order to increase resident awareness of telemetry status, residents were instructed to add a telemetry status column to their patient rounding lists in the electronic medical record (EPIC). This was followed by discussion of overuse of telemetry at an educational conference and distribution of telemetry guidelines in the conference handout. Statistical analysis was performed using χ² test.

Results: Only 13 % of residents checked orders for telemetry, 32% were not aware that the AHA telemetry guidelines existed and 61% were somewhat familiar with the guidelines. Prior to our interventions, 32% of patients were on telemetry, of which 67% were inappropriately on telemetry. After implementation of use of the telemetry column on resident Epic patient lists, 28% of patients were on telemetry, out of which 34 % did not have an appropriate indication. This resulted in 33% reduction in inappropriate use (p= 0.008). After discussion regarding overuse of telemetry at the program educational conference, 40% of patients were on telemetry, out of which 37% had inappropriate indications, resulting in 30% reduction of inappropriate usage from baseline (p= 0.012).

Conclusion: A significant proportion of general medicine patients are placed inappropriately on telemetry. This occurs despite institutional-level interventions to incorporate telemetry indications into the telemetry order. The physician culture of ordering telemetry as a safety net, lack of awareness of telemetry status, and limited knowledge regarding telemetry guidelines contribute to overuse. Our intervention to increase physician awareness of telemetry status through inclusion of the telemetry column on Epic patient list was effective in reducing inappropriate use. Subsequent resident education on telemetry guidelines was effective in helping to maintain a reduction in inappropriate telemetry orders but did not result in further reductions in inappropriate orders.
Introduction: The prevalence of obesity is rapidly increasing in the United States (US) and according to CDC, over 39% of adults in the US are obese. Studies have estimated that obesity increases national medical spending by over $150 billion each year. Large population studies from Denmark and the UK have demonstrated significantly higher rates of healthcare utilization by obese patients and also showed that hospital costs were almost 50% higher. However, the impact of obesity on outcomes in patients hospitalized with chronic pancreatitis has not been examined. We aimed to evaluate the impact of obesity on mortality and other outcomes in patients hospitalized with chronic pancreatitis.

Methods: A retrospective analysis was performed on cohorts of patients from the national inpatient sample database (NIS) from 2011-2014 and 2016. Patients over the age of 18 years admitted to hospital with a primary diagnosis of chronic pancreatitis were included in the study and outcomes in patients with obesity (BMI>30) were compared to those without obesity. The primary outcome was in-hospital mortality and secondary outcomes were healthcare resource utilization (length of stay, total hospitalization costs), number of abdominal magnetic resonance and computed tomographic imaging studies performed, major procedure undertaken including endoscopic retrograde cholangio-pancreatography (ERCP). Multivariate analysis was performed to identify independent predictors of in-hospital mortality after adjusting for confounders.

Results: A total of 102,458 patients were included in the study and 6,891 (6.7%) of these were found to be obese. After adjusting for confounders, obese patients with chronic pancreatitis had no difference in mortality as compared to non-obese patients [OR: 0.66, P-value: 0.580]. Obese patients, however, had increased length of hospital stay by an average of 0.5 days (95%CI: 0.10-0.93 days, P-value:0.015). Total hospital costs and charges appeared to be increased but could not reach statistical significance Rates of ERCP, abdominal imaging and need for total parenteral nutrition (TPN) were similar in both groups. Results are summarized in Table 1.

<table>
<thead>
<tr>
<th>Outcomes</th>
<th>Measures</th>
<th>P-Value</th>
<th>95% Confidence Interval</th>
</tr>
</thead>
<tbody>
<tr>
<td>In-hospital Mortality</td>
<td>aOR: 0.66</td>
<td>0.58</td>
<td>0.15-2.87</td>
</tr>
<tr>
<td>Mean Difference in LOS</td>
<td>0.51 days</td>
<td>0.015</td>
<td>0.10-0.93</td>
</tr>
<tr>
<td>Mean Difference in Total Charges</td>
<td>$4946</td>
<td>0.062</td>
<td>-252-10146</td>
</tr>
<tr>
<td>Mean Difference in Total Costs</td>
<td>$1031</td>
<td>0.10</td>
<td>-229-2292</td>
</tr>
<tr>
<td>Rate of ERCP</td>
<td>aOR: 1.00</td>
<td>0.966</td>
<td>0.72-1.40</td>
</tr>
<tr>
<td>TPN</td>
<td>aOR: 1.18</td>
<td>0.42</td>
<td>0.78-1.79</td>
</tr>
<tr>
<td>Abdominal Imaging (MRI/CT)</td>
<td>aOR: 0.83</td>
<td>0.47</td>
<td>0.49-1.38</td>
</tr>
</tbody>
</table>

Conclusion: Obesity increases length of hospital stay in patients admitted for chronic pancreatitis and it may possibly increase hospital costs. However, obesity does not have any impact of mortality in chronic pancreatitis and it does not have any impact on the need for ERCP, imaging or TPN. This study thus reinforces the impact of obesity on healthcare and highlights the need to be addressed urgently as the obesity epidemic continues to rage across the nation.
Diagnostic Accuracy of a Smartphone-Based Atrial Fibrillation Detection Algorithm

Authors: Isma Nusrat Javed MD, Nazir Ahmad MD, Stavros Stavrakis MD, PhD, David Albert MD

Introduction: Smartphone-based single-lead ECG devices have enhanced the feasibility of diagnosis and monitoring of arrhythmias, including atrial fibrillation (AF). The Kardia mobile ECG device is an FDA approved smartphone-based, single lead device, with an automated algorithm to detect AF, based on RR irregularity and absence of P waves. We examined the diagnostic accuracy of the Kardia Mobile algorithm for the diagnosis of AF in patients with paroxysmal AF.

Methods: Twenty-nine patients with paroxysmal AF and low CHADS2-VASc score were instructed to transmit a 30-second ECG every day and when experiencing symptoms for a median period of 20 months. The ECGs were transmitted to a secure server and the diagnosis was manually confirmed by 2 physicians. The sensitivity and specificity of the automated algorithm for the diagnosis of AF were compared against the physician interpretation as the gold standard.

Results: Over a median follow up of 20 months, 20 patients failed to submit a daily ECG at least once (median 3 failed submissions). A total of 14,998 ECGs were recorded. AF was diagnosed in 715 (5%) ECGs, while 1549 (10%) were deemed undetermined by the device. Overall, the kappa coefficient of agreement was 0.89 (95% confidence intervals 0.88 to 0.91; p<0.0001), indicating excellent agreement between the 2 methods. The device had a 99% sensitivity and 98% sensitivity for diagnosing AF. When the undetermined ECGs were treated as possible AF in the analysis, representing the worst case scenario, the specificity dropped to 87%, while the sensitivity was maintained at 99%.

Conclusion: The Kardia mobile ECG device provides excellent diagnostic accuracy in diagnosing AF, supporting the notion that such a device can be used for AF screening. In this setting, a high sensitivity in diagnosing AF will allow physicians to review only those recordings that are classified by the device as AF, in order to decrease the burden of having to review every transmitted ECG recording. The diagnostic accuracy of this single lead ECG device is critically dependent on high-quality signals. Thus, efforts should be directed toward patient education to acquire high-quality signals to optimize the performance of the device.
Improving Trends of Appropriate Aspirin Utilization for Primary Prevention of Atherosclerotic Cardiovascular Disease at A Tertiary Care Medicine Resident Clinic

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Introduction: Coronary artery disease (CAD) causes approximately one third of deaths in patients above the age of 35. Recent meta-analyses have demonstrated significant reduction in non-fatal MI and all-cause mortality with aspirin use for primary prevention strategies. We aimed to improve rates of appropriate aspirin treatment for primary prevention of CAD in our medicine resident clinic using a brief chart prompt.

Methods: Baseline rates of guideline-based aspirin use were established retrospectively from medical records of 200 randomly selected resident clinic patients aged 50-69. Those with known CAD, peripheral arterial disease, stroke, recent major bleeding, without a lipid profile within the previous 3 years or with an ASCVD risk < 10% were excluded. For the intervention arm, a 1-page prompt was placed in charts of consecutive patients aged 50-69 over a 1-month period. This prompt listed the USPSTF guideline and included four questions: 1) Should this patient be on ASA? 2) Is this patient on ASA? 3) Will you start ASA? and 4) If not, why? ASCVD risk and rates of aspirin prescription were confirmed by chart review. The proportion of patients treated with aspirin after institution of the prompt was compared to the baseline proportion using the two-proportion z-test. Also, prior to study initiation, we defined inappropriate ASA prescription based on the following criteria and assessed for inappropriate prescription in the intervention arm: history of fall, history of major bleeding, full dose anticoagulation use, and non-steroidal anti-inflammatory drug use.

Results: For the retrospective arm, 79 of 200 patients met inclusion criteria (48% male, 52% female, 98% hypertensive, 52% diabetic, and 70% previous or active smokers). Thirty-five of 79 (44.3%) were appropriately prescribed aspirin. Prompts were distributed for 80 patients in the intervention phase, and 26 met inclusion criteria. Of these, 19 (73.1%) were on ASA, yielding a 28.8% increase in rates of ASA use (95% CI = 6.7%, 45.6%, p=0.011). These results remained significant even after limiting the analysis to those aged 50-59 (rate increase of 31.1%, 95% CI 2.6%, 52.3%, p=0.03). Zero patients in the intervention arm were inappropriately prescribed ASA.

Conclusion: The USPSTF recommends prescribing aspirin for primary prevention of CAD among patients aged 50-59 with >10% 10-year risk ASCVD and a low bleeding risk. The benefit for patients aged 60-69 remains less clear; however, our patient population includes high rates of hypertension, diabetes, smoking, and despite appropriate statin therapy, elevated ASCVD risk. We feel that ASA for primary prevention is more beneficial than harmful in this population, when prescribed appropriately. Our short, simple questionnaire yielded significant improvement in rates of appropriate ASA prescription for primary prevention of ASCVD in the resident clinic setting.
Improving Recognition and Treatment of Depression in an Academic Internal Medicine Clinic

Authors: Ryan Yarnall MD, Summer Lepley DO, Stephanie Harry DO, Kavitha Mattaparthi MD, Kacey McConnell DO, Amy Wilson DO, Kristin Rodriguez MPH LSSGB, Carmen Vesbianu MD, Michael Weisz MD

Introduction: Many patients suffering from depression, one of the world’s leading causes of disability, are seen initially in primary care clinics, therefore, the recognition of and subsequent intervention for the disease are essential in these settings. We designed a quality improvement project with the aim of improving identification of depression by the provider with an appropriate follow up intervention.

Methods: In our clinic we screen every patient for depression yearly using the PHQ-2 tool. Our baseline data was collected from a sample of patients over the course of one month who had a positive PHQ-2. An individual chart review was performed looking for documentation of depression-related ICD codes and if any intervention was performed at the time of the visit. For the first PDSA cycle, we advised the nursing staff to review for positive PHQ-2 sheets and provide a brightly-colored PHQ-9 form to the patient for completion. The physician would theoretically notice these sheets and address depression during the visit. Our second PDSA cycle consisted of resident education on proper diagnosis and documentation of depression and guidelines-directed treatment with encouragement to schedule a 5 week follow-up if starting a medication. The third PDSA cycle consisted of educating social workers to contact patients two weeks after pharmacotherapy was initiated. Data end points were collected 2 months after these interventions were implemented.

Results: Before the intervention, 51.4% of patients seen in our clinic over the course of one month who screened positive on PHQ-2 had documentation of depression-related diagnoses. After the intervention, 68.4% of PHQ-2 positive patients had related documentation. The percentage of patients who had an intervention by the provider alone (medication prescription or adjustment with proper follow-up, referral to specialized mental health provider, patient-centered management) improved from 65.7% to 72.9%. The amount of the patients who received some sort of intervention, whether by a health care provider or social worker, remained relatively stable from 94.3% to 94.6%.

Conclusion: Our multi-disciplinary intervention appears to have improved provider performance in identifying and treating depression in the clinic environment. Future interventions and studies utilizing more specific depression identification tools including PHQ-9 are suggested.
Unlocking Implicit bias: Implementation of an implicit bias workshop to increase resident physician awareness of personal implicit bias and its effect on patient care.

Authors: Jacob Murray, DO; Vy Pham; Jabraan Pasha, MD, FACP, University of Oklahoma - Tulsa University School of Community Medicine, Department of Internal Medicine

Introduction: Unconscious attitudes, also known as implicit biases are ubiquitous and their effects are wide-ranging. From something as seemingly insignificant as the clutching of a purse in lieu of a passerby to something of potential great consequence such as the lack of a surgical referral, the fingerprints of implicit bias are on many of the decisions we make. However hard it is to conceive, physicians’ patient care decisions are not immune to the influence of unconscious beliefs and attitudes, and the data showing the effect of implicit bias on healthcare disparities is growing. Mirroring the societal disparities seen in the criminal justice system and employment, in healthcare, the divide is just as great. Given the strong data linking implicit bias to healthcare disparities, it is very important to promote awareness of implicit bias in not only healthcare, but also healthcare providers as individuals. Our study looked to increase resident awareness and confidence in managing unconscious attitudes and beliefs that may be affecting patient care.

Methods: Participants were 53 internal medicine, pediatric and family medicine residents at OU-TU School of Community Medicine. Between August 2018 and October 2018, during Academic Grand Rounds, participants attended a 90-minute implicit bias workshop. The interactive workshop looked to define implicit bias, demonstrate its origins and show its societal impact via lecture, hands-on activities and small group discussion. Via a 7-point Likert scale, a pre and post-workshop questionnaire measured participants’ awareness of implicit bias and confidence in discovering and managing implicit attitudes. Data was analyzed via paired T-test analysis.

Results: There was a mean increase of resident physicians’ awareness of general, personal implicit bias (mean increase of 0.938, P<.001) and awareness of implicit bias toward patients (mean increase 0.698, P<.001). The results also reflected an increase in confidence in discovering personal implicit bias (mean increase 0.358, P<.001) and in management of implicit attitudes discovered about patients (mean increase 0.321, P<.001).

Conclusion: Our implicit bias workshop proved successful in increasing resident awareness of their vulnerability to unconscious attitudes and in increasing their confidence in recognizing and managing these same attitudes. Although increasing awareness, alone is not an adequate strategy for eliminating implicit bias, it has been shown to be effective in beginning to limit unconscious bias. Paired with increased confidence in recognizing and managing these biases, this workshop appears to be a practical and effective first step towards combatting implicit bias.
Frailty and cardiovascular mortality: a pooled analysis of individual patient data

Authors: Malik Farooqi1,2, John Liu2, Leanne Dyal2, Salim Yusuf2, Darryl Leong2,3, 1Department of Medicine, McMaster University, Hamilton, Canada, 2Population Health Research Institute, McMaster University and Hamilton Health Sciences, Hamilton, Canada, 3Department of Health Research Methods, Evidence, and Impact, McMaster University, Canada

Introduction: Frailty is associated with higher mortality in individuals at high cardiovascular disease (CVD) risk. It is unclear if frail patients are at high-CVD risk or if they die from competing non-CVD causes. Our objective is to understand how frailty leads to premature mortality in patients with or at high risk for CVD, by evaluating the relationship between frailty, and incident CVD, CVD death and non-CVD death.

Methods: We conducted an individual patient pooled analysis of patients with, or at risk for CVD, recruited to 14 multicentre randomized controlled trials. Frailty was determined by the cumulative deficit model (frailty index) using baseline data. Our primary outcomes were 1) incident CVD (defined as new MI, new stroke or new heart failure), 2) mortality (CVD and non-CVD) and 3) CVD case-fatality. Incident CVD was evaluated by calculating standardized incidence rate ratios (IRR) using Poisson regression. The relationship between frailty and the mortality was determined by calculating hazard ratio’s (HR) and 95% confidence intervals (CI) using time-to-event models based on the Cox proportional hazards assumption as well as Kaplan-Meier curves. To mitigate within-study clustering, we used shared frailty models in our time-to-event analysis, in which the study was modeled as a random effect. All analyses were age and sex-adjusted.

Results: We studied 155,270 patients (mean age 70.8 years, 63% male) with median follow-up of 3.2 years, of whom 37% were frail. Frailty was associated with a higher risk of incident CVD compared to non-frailty (incident rate ratio 3.10, 95% CI 2.91-2.31). Frailty was also associated with a higher all-cause mortality (hazard ratio, HR 2.06, 95% CI 1.91-2.22), CVD mortality (HR 2.54, 95% CI 2.30-2.81) and non-CVD mortality (HR 1.52, 95% CI 1.36-1.70) than non-frailty. Following an incident CVD event, the case-fatality rate was higher in frail (35.1%, 95% CI 33.9-36.2%) than in non-frail (20.0%, 95% CI 17.8-22.4%).

Conclusion: In individuals with or at high risk of developing CVD, frailty portends a poorer outcome in different ways, including enhancing the risk of incident CVD, increasing the risk of death if CVD develops, and both CVD and non-CVD mortality. Therefore, targeting frailty may represent an important approach to improve CVD outcomes.

References

HEART Score as a Risk Stratification Tool For Patients With Chest Pain at a Community Emergency Department

Authors: Mark Day, DO, Sally Mangum, PhD, DO, Olivia Pipitone MPH, and Jesse Greenblatt, MPH, MD

Introduction: Acute coronary syndrome (ACS) includes unstable angina, non-ST elevation myocardial infarction, and ST elevation myocardial infarction. Risk factors for ACS include hypertension, hyperlipidemia, diabetes, and other common comorbidities. The HEART score is a risk calculator that was developed in 2008 to determine the risk of major adverse cardiac events within 6 weeks of presentation to the ED with chest pain. HEART scores estimate patient risk for major adverse cardiac events within 6 weeks. It uses history, electrocardiogram, age, risk factors, and troponin.

Methods: Retrospective cohort study using EHR data. In a previous study, we demonstrated that 14% (n=249) of adults who presented with chest pain to the ED of a community hospital in suburban Oregon in 2016 were eventually diagnosed with ACS. For the present study, demographic data and data on risk factors for ACS were collected on the 249 patients who had ultimately been diagnosed with ACS. Chart review was performed to fill in additional information on gender, age, ECG, troponin, and risk factors including: DM, HLD, HTN, Smoking status, personal and family history of ACS, and Obesity. HEART Scores were calculated for each patient.

Results: Results showed a high burden of hypertension (78%), hyperlipidemia (64%), and an overall high burden of our measured risk factors (98% had at least one risk factor). Average patient HEART Score was 6.3, which corresponds to a moderate to high risk of a major adverse cardiac event within 6 weeks. 111 individuals (45%) were in the high risk category for HEART Scores, 130 (52%) were moderate risk, and 8 (3%) were low risk. Across all HEART Score groups, a majority of patients were diagnosed on the same day as their ED encounter for chest pain (75%-81%) or within 6 weeks of the ED encounter (14%-25%). 51 patients were diagnosed with ACS more than 24 hours after presentation. However, at the time of their presentation to the ED, 49 of them had moderate or high risk HEART Scores (96%).

Conclusion: Results showed a higher burden of hypertension (78%) and hyperlipidemia (64%) in this population than in the general population. Studies have estimated the prevalence of hypertension to be between 25% to 38% and the prevalence of LDL to be between 20% and 27% in the general population. 49 patients who were not diagnosed with ACS at their initial ED encounter had a moderate or high HEART Score at the time of their ED encounter. With utilization of HEART Score, these patients would likely have had further workup and possibly earlier detection. These findings elucidate prevalent risk factors and the functionality of the HEART Score in our community ED for adults who present with chest pain.
"Just" a phone call away: Reducing 30 day Readmission Rates through Increased Outpatient Follow Up

Authors: Cody Talbot, DO, Graham Wolf, MD, Shelley Sanders, MD

Introduction: Hospital readmission within 30 days of discharge is a major burden on the US Healthcare system. Readmissions not only indicate a poor prognosis but they also negatively impact hospitals by lowering Medicare reimbursement. At our 523-bed community teaching hospital, the 2015 all-cause readmission rate was 10.5%, increased from 2014. This resulted in a 0.03% reduction in reimbursement through the CMS Hospital Readmissions Reduction Program. This quality improvement project aimed to increase out-patient follow-up within 14 days of discharge from one hospital unit in order to reduce 30-day readmission.

Methods: The project was initially designed as a prospective, crossover study between two resident ward teams. Any Providence Medical Group patient admitted to the Accountable Care Unit (ACU) was enrolled into the study. Once ready for discharge, residents were instructed to prompt health unit coordinators (HUCs) to schedule outpatient follow-up utilizing a protocol within 14 days. This initial pilot occurred from 10/16/17 through 12/10/17. Unfortunately, the protocol proved to be too complicated, requiring dependence on generating inpatient lists to identify the study group. Crossover also occurred between intervention and control teams in utilizing the HUCs for discharge. Based on these findings, the protocol was simplified in several ways. First, all patients were included rather than just one subset. Second, hospitalist teams managing patients on ACU were enrolled as control group while both resident teams represented the intervention arm. Finally, a HUC appointment log was created to act as a process measure. The study relaunched on 03/05/18 and concluded on 7/29/18.

Results: After the relaunch, 329 patients were discharged and 66 were readmitted within 30 days. Unfortunately, the intervention did not appear to increase outpatient follow-up within 14 days. For interest, we assessed readmission rates among patients who did and did not get 14 day follow-up. Data revealed only 15.6% of patients were readmitted when 14 day follow-up occurred, while 24.2% of patients without 14 day follow-up were readmitted (p=0.0548).

Conclusion: Despite two cycles of change, increasing 14-day follow-up proved difficult; however, failure mode and effects analysis identified several root causes. First, review of the process measure suggests crossover occurred between intervention and control groups in utilizing the discharge protocol again. Second, the EMR query used to identify whether patients had follow-up was only able to capture a subset of patients, as some providers don’t use our health record. Finally, HUCs often failed to secure an appointment due to lengthy hold times, suggesting that our default practice of leaving this task to a recently ill patient or family member is dubious at best. Electronic scheduling or an outpatient hospital discharge clinic operated by hospitalists may represent simpler solutions.
**OREGON RESEARCH POSTER FINALIST - QIAN ZHOU, MD**

**Improving Hepatitis B Vaccination Rates in Diabetic Patients**

Authors: Qian Zhou, MD; Shelley Schoepflin Sanders, MD; Irene Hendrickson, MD; Alyssa Nelson

**Introduction:** Chronic hepatitis B Virus (HBV) infection causes cirrhosis and hepatocellular carcinoma in 20-30% of affected patients. Diabetic patients have a higher risk for acute HBV than non-diabetics due to regular skin breaks and are more likely to remain chronically infected. The Advisory Committee on Immunization Practices recommends hepatitis B vaccination for all adults aged 19-59 with diabetes (Type I and II), but in ambulatory practices, vaccination rates are often low. This multi-intervention quality improvement project aimed to improve hepatitis B vaccination among diabetic patients in a residency teaching clinic using a mailed letter campaign and visual vaccine reminders at office visits.

**Methods:** The baseline vaccination rate among diabetics aged 19-59 in a residency teaching clinic was measured at 22%. All patients aged 19 to 59 who were due for ≥1 hepatitis B vaccines received a letter written in plain language which included a vaccination reminder, a CDC guideline for hepatitis B immunization for diabetic patients, and a timetable indicating when their next vaccination was due. When eligible patients arrived, Clinic Care Coordinators placed a laminated vaccination reminder in the room to prompt providers. In addition, clinicians and residents completed two education sessions. Pre- and post-intervention data were obtained through Epic EHR for the intervention clinic and a similar control clinic.

**Results:** 164 eligible patients were identified and sent a letter; the control clinic had 174 eligible patients. The return-rate was 21 patients vaccinated out of 164 letters sent (12.8%). Between November 2017 and September 2018, the percentage of patients with any hepatitis B immunization increased from 19% to 31% in the intervention clinic; the control clinic was unchanged (35% to 36%. P=0.0001). In the intervention clinic, the percentage of patients receiving all 3 hepatitis B vaccinations increased from 12% to 19%; the control clinic was stable (23% and 25%. P=0.0159).

**Conclusion:** Education, provider reminders, and patient engagement through letter outreach cost effectively improved hepatitis B vaccination significantly over 11 months. The comparison clinic has an on-site endocrinologist, which could explain the higher vaccination rate at baseline, and illustrates the importance of patient and clinician education. One limitation of the intervention is patient accessibility by mail or phone. The follow-up period is only 11 months; a longer study might better assess the impact of the intervention as more patients could complete all 3 vaccinations. It is unclear if multiple reminders or more education will further improve the vaccination rate, and further investigation might clarify these questions.

**References**

Predicting Major Adverse Cardiac Events using HEART Score in the Elderly

Authors: Kevin Gu MD, Amar Kaneria MD, Parija Sharedalal MD, David Chang MD, Neel J Patel MD, Kevin D’Mello MD, Andrew Kohut MD

Introduction: The HEART score was developed to predict Major Adverse Cardiac Events (MACE) within 6 weeks of being evaluated for acute coronary syndrome. In the established score, age is scored as <45 years old (y/o) = 0, 45-64 y/o = 1, and >65 y/o = 2. The average age of patients in the pilot study was 61.2 y/o. Our aim is to evaluate the predictive value of the HEART score in patients >65 y/o. We hypothesize that elderly patients are at increased risk of MACE unaccounted for by the established age scoring system.

Methods: We retrospectively reviewed 668 emergency room visits for chest pain (exclude STEMI) between 2016 to 2018. HEART scores were calculated based on presenting data. The primary outcome was MACE (MI, PCI, CABG, or death) within 6 weeks. Primary outcomes were compared between various age groups using the established age scoring system and additional stratification of patients >65 y/o. Differences in HEART scores were controlled by comparing patients within the same HEART score brackets (4-6, 7-8, 9-10). Chi squared analysis was used to determine statistically significant differences in MACE (p<0.01).

Results: The average age was 60.2 y/o and average HEART score 5.56. There was statistically significant increase in MACE between <45 y/o (18.5%), 45-64 y/o (36.6%), and >65 y/o (47.4%) as outlined by the established HEART score (p<0.01). However, when comparing 65-74 y/o (45.7%), 75-84 y/o (48.8%) and >85 y/o (48.8%), increasing age did not correlate with increasing rate of MACE (p=0.90). After controlling for differences in HEART scores, there was again no statistically significant increase rates of MACE across all HEART score brackets.

Conclusion: In patients >65 y/o, increasing age did not correlate with increasing rate of MACE. Our findings support the current age stratification of the established HEART score and show that even in the very elderly, the HEART score adequately accounts for age as a contributor to MACE. We plan to further define our results using logistic regression analysis of individual HEART score components to determine if other components of HEART score have higher predictive value in this population.
Trends in hospitalizations related to anaphylaxis, angioedema and urticaria in the United States (2001-2014)

Methods: Using the largest inpatient National Inpatient Sample data in US from 2001-2014, all age groups admitted with a primary diagnosis of anaphylaxis, angioedema, urticaria were identified based on International Classification of disease-9 codes. Yearly distribution of hospital admissions was stratified per different age groups. STATA version 13.0 (College Station, TX), MS-Excel 2016 and Joinpoint Regression Program version 4.5.0.1 were used to determine yearly trend of hospitalizations related to anaphylaxis, angioedema and urticaria.

Results: While an increasing trend in the rate of hospitalizations was seen for angioedema (annual percentage change [APC] 4.48), a decreasing trend (APC -2.19) was observed for urticaria-related hospitalizations. Overall anaphylaxis-related hospitalizations were noted to be stable, but a significant increasing trend was observed among age group 5-14 years (APC 4.19), mostly due to the subgroup of food-related hospitalizations (APC 5.86). Angioedema-related hospitalizations were found to be highest among the 35-64 years age group (APC 5.38).

Conclusion: An increasing trend of hospitalizations has been observed for allergic conditions, with varying age-distribution according to the nature of eliciting agent and susceptibility of different age groups. While angioedema has been observed as an increasing problem in older population, food-induced anaphylaxis is a growing concern in the younger population.

References


Improving the Efficacy of Chemotherapeutic Agents Through the Use of Stapled Peptide Mimetics

Authors: Amogh Joshi DO, Jan Jezek PHD, Randy Strich PHD

Introduction: The mitochondria serve as a key regulatory center for stress-induced programmed cell death (PCD). Mitochondrial fission serves as the preliminary step in this process. Induction of PCD through oxidative stress is a common mechanism for anti-neoplastic agents. Cyclin C, a ubiquitous transcription factor, has been recently shown to translocate from the nucleus to the cytoplasm in response to stress. Previous studies in yeast have determined that deleting the nuclear cyclin C anchor, Med13p, releases cyclin C into the cytoplasm where it induces mitochondrial fission and potentiates cellular hypersensitivity to oxidative stress. Here we hypothesize that manipulation of cyclin C localization could increase sensitivity of cancer cells to reactive oxygen species (ROS)-mediated PCD. Hence, we aimed to disrupt the cyclin C – Med13 interaction by the addition of different stapled peptide mimetics (S-HAD) designed to target the holoenzyme activating domain (HAD) of cyclin C.

Methods:

Mitochondrial Fragmentation Assay - Cells were cultured on coverslips for 2 days, treated with the indicated concentrations of S-HAD for 2 hr, stained with 100 nM MitoTracker Red CMXRs (Molecular Probes, Grand Island, NY) for 30 min, and fixed with 4% paraformaldehyde for 10 min, permeabilized with 0.2% Triton X-100 for 10 min, blocked with 2% BSA, and incubated with 1:250 cyclin C (PA5-16227, Thermo Fisher Scientific) and 1:2,000 AlexaFluor® 488 conjugated secondary (A11008, Thermo Fisher Scientific) antibody (overnight and 1 hr respectively). Fixed cells were mounted with 4′,6-diamidino-2-phenylindole (DAPI)–containing medium to stain the nuclei and the images were acquired with a Nikon Eclipse 90i epifluorescent microscope (Melville, NY) equipped with a Retiga Exi charge-coupled device camera. Representative images were taken with the 100×objective, while mitochondrial fragmentation was quantified using the 60×objective. Cells containing 15 or more mitochondrial puncta were considered fragmented.

Confocal Live Cell Imaging – Cells were grown on Petri dishes for 1 day and stained with MitoTracker Red. The cells were imaged on the Nikon Eclipse C1Ti confocal microscope.

Annexin V Assay – Annexin V-FITC flow cytometry assays were conducted as described by the manufacturer.

Statistical Analysis - Σn refers to the number of samples taken, N refers to the number of independent experiments. P-values were determined using the Student’s t-test.

Results: S-HAD treatment induced partial cyclin C release from the nucleus and showed significant and dose-dependent mitochondrial fragmentation in wild-type but not cyclin C null mouse embryonic fibroblasts. Similarly, S-HAD3-treated HeLa and MCF-7 cells showed a fragmented phenotype. Moreover, S-HAD3 sensitized HeLa and MCF-7 cells to cisplatin treatment as assessed by Annexin V assay.

Conclusion: In conclusion, these results indicate that manipulating cyclin C localization with stapled peptides effectively induces mitochondrial fission and sensitizes tumor cells to conventional chemotherapeutics.
References

Introduction of continuous EEG (cEEG) at a community teaching hospital enhances care and leads to a reduction of hospital to hospital to transfers

Authors: Ammar Malik, MD; Asif Abdul Hameed, MD; Kakageldi Hommadov, MD; Sonul Gulati, DO; Philip Choi, MD; Dominic J. Valentino, DO

Introduction: Continuous EEG offers crucial information which can drastically change a patient’s outcome. Until recently, the cost of implementation was prohibitive and limited to tertiary care centers with Neuro-ICUs. Recent technological advances have reduced the cost and remote access has made the interpretation of results feasible by alleviating hospitals from having an on-call epileptologist. Our community teaching hospital would previously transfer all patients suspected of status epileptics/seizure disorders to tertiary care centers for cEEG monitoring. This would lead to discontinuity of care, added handoffs and potential for destabilization during transit. Additionally, there is a loss of operating revenue to the hospital as these patients often carry additional comorbidities and contribute significantly to the ICU’s case mix index (CMI), which affects reimbursements from CMS.

Methods: A retrospective chart review was completed of all patients who were placed on cEEG during its first year of implementation. Patients were admitted from the emergency department or the medical floors. Patients were followed from admission to discharge. We tracked patient outcomes including ICU LOS, days on cEEG and discharge disposition.

Results: In just the first year of implementation, 26 patients were placed on cEEG and a total of 28 studies were performed. Of these, 17 patients were admitted with an initial diagnosis of seizures; the remaining 10 were admitted with a different diagnosis but seizures were suspected after admission to the ICU. Five were diagnosed with status epilepticus. Only two of these patients were transferred to a tertiary care center with Neuro-ICU for uncontrolled seizures. One of these patients had a tumor which required special neurosurgical intervention. 10 patients were found to have findings consistent with moderate to severe encephalopathy. 12 patients were ruled out for seizures and were either discharged home or to a SNF. The average time on cEEG per patient was 2.3 days

Conclusion: cEEG in the ICU is an invaluable tool that can now be utilized cost effectively in the community hospital setting. It allows rapid diagnosis and treatment of seizures which can improve patient outcomes. This results in reduction of hospital to hospital transfers. Simultaneously, it improves the ability of hospitals to maintain a higher CMI in their ICU, with potential to cover the costs of the cEEG program. Additionally, retaining these patients also allowed our house-staff to learn about the management of seizures in a more in-depth fashion. We believe this is a cost-effective diagnostic intervention which can now be safely and effectively used at community teaching hospitals such as ours.
A Study of Simple Syncope- Are we Overtesting?

Authors: Muhammad Hassaan Sattar; MD; Muhammad Usman Ali, MD; Christopher struby, DO; Mirnove Domond, DO; Jugeet Kanwal, MD; Neha Puri, MD, Khsitij Thakur, MD, Jonathan Finkle, MD

Introduction: Syncope accounts for 1-6% of hospital admissions and 1-3% of emergency room visits per year resulting in significant medical costs. A Simple Syncope is defined as syncope without neurological deficit or seizure. Although computed tomography, carotid ultrasound and echocardiogram is not indicated in patients with simple syncope, millions of patients get these tests done unnecessarily. This strategy rarely identifies an underlying cause and is not cost effective. We performed a retrospective cohort study examining the patients with simple syncope to determine the use of imaging according to high value care guidelines.

Methods: A retrospective chart review was performed on a total of 630 patients who presented to the Emergency Department at an academic institution with initial diagnosis of syncope over the previous 4 years (2014 –2017). Information collected include determination of simple syncope; if CT head, carotid ultrasound and echocardiogram was done with proper indications; if orthostatic vitals were checked and San Francisco Syncope (SFS) score was calculated.

Results: Of the 630 patients reviewed, 361/630 (57%) patients were identified as having a simple syncope episode and 66% (239/361) of them underwent CT scan of head to rule out intracranial pathology. The number of CT head done by the ED was 92% (221/239), 2% (4/239) was done by the medical admitting resident and 6% (14/239) was done by the primary team. Interestingly 96% of these CT Scans did not show any pathology and only 3% (8/239) showed TIA/stroke and 1% (2/239) showed brain lesion or bleed.

In our study 16% (58/361) of the patients admitted for simple syncope had carotid ultrasound done during the hospital stay and 97% (57/58) of them did not show any significant stenosis. We also found that 39% (141/361) of these had echocardiogram done and 93% (132/141) of them did not show significant abnormality.

In terms of orthostatic vitals, only 32% (116/361) of patients had their orthostatic vitals checked, out of which 6% (8/116) were done by ER, 1% (1/116) by the admitting resident and 25% (30/116) by the primary team. A series of Chi-square tests showed there were no significant differences in carotid ultrasound, and echocardiogram between teaching and hospitalist services (P-value for echo=0.4309, P-value of USG=0.3336).

For risk stratification, the patients with higher SFS score had a higher average in-hospital stay compared to low SFS score.

Conclusion: Syncope is one of the most frequent presenting complaints in Emergency Room and patients need to be stratified based on risk factors. Despite evidence and guidelines, we found that we are doing more testing that is inconsistent with high value care guidelines. The recognition of inappropriate use of imaging in patients with simple syncope can help significantly reduce healthcare expenditures. Utilizing a standardized approach to syncope evaluation can help.
Motivations behind Influenza Vaccination in Hispanic Patients with Influenza Infection during the 2017-2018 season.

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Introduction: Despite the availability of anti-viral therapy, vaccination remains a cornerstone in the prevention of influenza infection. However, vaccination rates in the United States remain low. A report by the CDC placed the influenza vaccine coverage for the last season around 37% for adults, a fall of 6.2% from last season.[1] Recently, Boey L, et al published data where they describe the positive and negative factors that affect vaccination uptake among HCW in Belgium.[2] However, data on influencing factors for influenza vaccination among the Hispanic population is limited. In this study, we interviewed a group of Puerto Rican patients infected with the influenza virus during the season of 2017–2018. The participants were interviewed via telephone and motivations to undergo or to forgo vaccination were explored. We aimed to uncover the forces that drive the decision for vaccination among this population.

Methods: This study was designed as a cross-sectional study. The data was retrieved from a community hospital in the northern region of Puerto Rico. Non-pregnant adults with a positive rapid influenza test during the 2017-2018 influenza season were included in the study. A questionnaire composed of a section to assess their knowledge of the influenza infection and another on intentions and motivations for vaccination was constructed. Participants were called via telephone and after consent, they were interviewed. Clinical and demographic data was retrieved from the electronic health record.

Results: A total of 50 participants were interviewed, representing a response rate of 33.1% (50/151), and most of them were females 37/50 (74%). Most of the participants were not admitted to a ward leading to an admission rate of 18% (9/50). A small proportion of the sample was vaccinated 11/50 (22%). In this group, the main motivation for vaccination was "being recommended by a physician" 5/11 (45.5%) followed by "being provided at work" 2/11 (18.2%). Those who were not covered prior to the event represent a 39/50 (78%) of the sample. The main reason for avoiding vaccination was trust issues 11/39 (28.2%), followed by unspecified reasons 10/39 (25.6%), and lack of time 7/50 (17.9%). In the knowledge section 43/50 (86%) answered that the virus can be spread from person-to-person, 42/50 (84%) responded that influenza can be a cause of death and 45/50 (90%) acknowledge that it can cause severe complications.

Conclusion: Trust issues regarding the influenza vaccine seem to be an important factor to forgo vaccination. However, accessibility of vaccine in the work setting and a recommendation by a physician seems to be a positive factor that motivates vaccination. Despite being well aware that this infection can be a cause of death and of severe complications the vaccination rate remained lower than most of the population in the United States. This information can help the shape of further vaccination campaigns.

References

Frailty Increases the Risk of Non-Home Discharge and Hospital Length of Stay in Elderly Patients Undergoing Elective Major Surgery

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Introduction: Surgical procedures amongst the elderly are increasing, but these patients’ postoperative trajectory is still difficult to predict. Frailty has been associated with higher risk of mortality and postoperative complications. Few studies have examined the relationship between frailty and disposition at discharge or hospital length of stay.

Methods: We conducted a prospective cohort study in a single academic center between January 2017 and 2018. We recruited patients of 65 years or older undergoing major elective surgery. We administered the Clinical Frailty Scale (CFS) during preoperative evaluation and patients were classified as robust, prefrail or frail.

A research assistant blinded to their frailty state collected baseline data and postoperative outcomes through chart review. Patients could either return to their primary residence or be discharged to a nursing home, a rehabilitation center or a long-term care center. We performed multivariate logistic regression and negative binomial regression to evaluate respectively the association between CFS and disposition at discharge as well as its association with hospital length of stay.

Results: A total of 271 patients were included. The median age was 72.0 years old and the median Charlson Comorbidity Index was 4.0. Based on the CFS, 50.9% of patients were robust, 32.5% were prefrail and 16.6% were frail. Patients underwent elective orthopedic surgery (61.6%), general surgery (24.7%) or vascular surgery (13.7%).

After adjusting for age, comorbidity and ASA score, being prefrail increased the odds of non-home discharge by 6.0 (95% CI 1.8 - 20.8), while being frail increased the odds by 16.2 (95% CI 4.5 - 58.4). The addition of CFS to age, comorbidity and ASA score increased the AUC from 0.74 to 0.83. Frail patients also had a longer hospital length of stay compared to robust patients (incidence risk ratio 1.7; CI 1.2 - 2.4).

Conclusion: The Clinical Frailty Scale predicted adverse outcomes in our cohort of elderly surgical patients. It is a simple tool that could be easily integrated in patient management before and during hospital stay to identify patients at higher risk of postoperative complications.
Left Atrial Enlargement is Independently Associated with Cardioembolic Stroke and Detection of Atrial Fibrillation after Embolic Stroke of Unknown Source

Authors: Kevin Jordan, Shadi Yaghi, Athena Poppas, Andrew Chang, Brian Mac Grory, Shawna Cutting, Tina Burton, Mahesh Jayaraman, Alexander Merkler, Hooman Kamel, Mitchell S. V. Elkind, Karen Furie, Christopher Song

Introduction: Left atrial enlargement has been shown to be associated with ischemic stroke but the association with embolic stroke mechanisms remains unknown. We hypothesize that increased left atrial volume index (LAVI) is more prevalent among patients with cardioembolic stroke than other stroke subtypes, and predicts AF detection on cardiac event monitoring in patients with embolic stroke of unknown source.

Methods: Data was collected from a prospective cohort of ischemic stroke patients admitted to a single academic center during an 18-month period. Stroke subtype was classified into cardioembolic stroke (CES), non-cardioembolic stroke of determined mechanism (NCE), or Embolic Stroke of Undetermined Source (ESUS). To assess the association between LAVI and atrial fibrillation (AF) detection in patients with ESUS, we included all patients with ESUS who underwent transthoracic echocardiography (TTE) and outpatient cardiac event monitoring. Comparison was made using multivariate logistic regression models. To assess the association of LAVI with stroke subtype we compared CE vs. NCE stroke and ESUS vs. NCE stroke using multivariate logistic regression models.

Results: Of 1234 patients identified during the study period, 1020 (82.6%) underwent TTE at time of ischemic stroke and had LAVI measurements. Stroke subtypes were: 336 (32.9%) CES, 412 (40.4%) ESUS, and 272 (26.7%) NCE. LAVI was greater in patients with CES than NCE (41.4 mL/m² ± 18.0 versus 28.6 mL/m² ± 12.2, p < 0.001). There was no difference in LAVI between patients with ESUS vs. NCE (28.9 mL/m² ± 12.6 vs. 28.6 mL/m² ± 12.2, p =0.61). Fully adjusted multivariate logistic regression models demonstrated that LAVI was greater in CE vs. ESUS (adjusted OR 1.07, 95% CI 1.05-1.09, p < 0.001), but not significant different in LAVI when comparing NCE vs. ESUS (adjusted OR 1.00, 95% CI 0.99-1.02, p = 0.720). Among 99 patients with ESUS who underwent cardiac monitoring, 18.2% had AF detected; LAVI was independently associated with AF detection in ESUS (adjusted OR 1.09, 95% CI 1.02-1.15, p = 0.007).

Conclusion: LAVI is associated with cardioembolic stroke as well as AF detection in patients with ESUS, two subsets of ischemic stroke which benefit from anticoagulation therapy. Patients with increased LAVI may be a subgroup where anticoagulation may be tested for stroke prevention.
Bongs and Barotrauma. Inhaled marijuana and spontaneous pneumomediastinum, a retrospective review

Authors: Zoe Weiss MD, Sara Gore MD, Andrew Foderaro MD

Introduction: Inhaled marijuana is infrequently reported as a potential risk factor for the development of spontaneous pneumomediastinum (SPM), a rare finding of extraluminal gas in the mediastinum, thought to be caused by barotrauma during breathing maneuvers. Other risk factors include coughing, vomiting, weight-lifting, childbirth or other forceful straining. Patients may present with severe chest or neck pain and subcutaneous crepitus. Though SPM is typically self limiting, patients are frequently admitted for observation and workup to exclude esophageal injury. The mechanism by which patients inhale marijuana is often not ascertained by physicians, thus little is known about how different smoking techniques or breathing maneuvers precipitate pulmonary injury. We aimed to evaluate the frequency of marijuana use in patients with SPM over a 12-month period and determine the extent to which clinicians document smoking histories.

Methods: We performed a retrospective chart review examining patients presenting to the hospital with a diagnosis of pneumomediastinum at a 719 bed urban tertiary care center between January-December of 2016. Cases were excluded if pneumomediastinum was precipitated by trauma, malignancy, or iatrogenic cause. The remaining cases were deemed to be "spontaneous" and were reviewed for reports of inhalational marijuana. The mechanism and frequency of smoking were documented along with the temporal relationship between smoking and of symptoms if elicited by provider. Co-existing risk factors, vital signs, presenting symptoms, and clinical management were also documented.

Results: Out of 147 cases of pneumomediastinum, 21 were classified as spontaneous, of which 14 (66.7%) were associated with marijuana use. Average age was 22.5 years (range 18-30), with male predominance (64.2%). Daily or more use was reported in 50% of cases. Three patients specifically reported symptoms onset during or immediately after smoking marijuana. The mechanism of smoking was described in only two cases. One patient reported using a “large bong” and another a “face mask.” Concurrent risk factors such as vomiting (57.1%) and coughing (42.9%) were commonly present. Counseling was provided in 50% of cases.

Conclusion: Inhaled marijuana is likely an underappreciated risk factor for the development of pneumomediastinum, likely due to barotrauma during strained breathing maneuvers such as forcible inspiration against a closed airway or forcible exhalation (or breath holding) against a closed glottis. Forced inhalation through a high-resistance smoking apparatus, such as a bong, can cause a drop in intrathoracic and peri-alveolar pressure, generating a transmural pressure gradient between alveoli and their vascular structures, leading to shearing damage and air leakage into the interstitium around the bronchovascular sheaths (Macklin phenomenon) into the mediastinal structures. The frequent presence of co-existing risk factors including coughing or vomiting suggests a possible additive effect. Given current advances in the legalization of recreational marijuana and the increasing use of medical marijuana, further investigation into the risks of specific smoking maneuvers and delivery devices is necessary. Physicians should be aware of the association between SPM and inhaled marijuana in order to provide appropriate counseling.

References

AN ADAPTIVE APPROACH TO CURBING TELEMETRY OVERUSE

Authors: Jessica L. Schachter, PGY-2, DO and Paari Gopalakrishnan, MD

Introduction: The overuse of telemetry produces a significant cost burden, facilitates inappropriate testing for rhythm artifacts, and contributes to a shortage of telemetry beds. Reducing the use of telemetry has not demonstrated a negative impact to overall patient outcomes. Our objective was to improve appropriate application and reduce duration of telemetry as part of a greater goal to preserve healthcare resources.

Methods: Over the course of 13 months, we initiated a protocol aimed at improving utilization of cardiac monitoring. The protocol was derived from the updated 2004 American College of Cardiology and American Heart Association guidelines. The protocol involved three phases. In phase I, telemetry orders would newly require designation of 24-hour or continuous monitoring with suggested indications for each. In phase II, an additional option of 48-hour monitoring was added with refinement of these indications. In phase III, efforts were targeted at nursing education on prompt discontinuation of telemetry per protocol. Statistical analysis was completed through utilization of the Applied Analysis of Variance (ANOVA) to test the equality of total telemetry usage, in hours, per day across all three phases. Pairwise test with Bonferroni-corrected p-values followed.

Results: From phase I to phase II, there was an increase in telemetry-hours (3147 vs 3375 hours) of 0.834. From phase I to phase III, there was a decrease in telemetry hours (3147 vs 2601 hours) with a corrected p-value of 0.078. From phase II to phase III, there was a significant decrease in telemetry hours (3375 vs 2601) with a corrected p-value of 0.009.

Conclusion: Consistent with prior studies, we demonstrated an improvement in telemetry overuse after implementing a guideline-based protocol. Notably, improvement in average length of telemetry monitoring was not observed until the application of directed nursing education. Further improvements might be directed toward physician and nursing education.
SOUTH CAROLINA RESEARCH POSTER FINALIST - YUAN YAO, MD

ARE YOU MY DOCTOR? ASSESS AND IMPROVE PATIENT IDENTIFICATION OF PHYSICIANS AND THEIR ROLES BY DISTRIBUTING INFORMATION CARDS

Authors: Yuan Yao, MD, Savannah Desmarais, MD, Frank Weigel, DO, Payal Parikh, MD, Victor Collier, MD, Nino Balanchivadze, MD

Introduction: Multiple medical team members and trainees are involved in direct patient care at a teaching institution leading to confusion with regards to whom is ultimately responsible for patients care. At times this may result in an unclear delivery of the plan of care to patients and their families and impair their ability to make informed decisions. Prior studies have shown that most patients (75-87%) were unable to identify an inpatient physician in charge of their care. Studies suggest placement of cards listing names, photos, and roles of team members in patients’ rooms significantly improved patients’ ability to identify their in-hospital physicians in a teaching hospital. However, patient satisfaction was not evaluated in these studies. The purpose of this QI project is to distribute information cards to improve patients’ identification of their in-hospital physicians and understanding of physician team and ultimately improve communication and patient satisfaction at our institution.

Methods: This quality improvement project included a pre-information card phase and an information card phase. A feedback sheet was administrated in both pre-information card phase and information card phase to all eligible patients. Survey results were assessed at regular intervals. An information card was developed based on a prior study, which included each physician’s name and their roles in a teaching team. The intervention was to distribute an information card to all patients on the internal medicine teaching team. These cards were updated weekly when team members changed. Patient satisfaction information was collected as very satisfied, somewhat satisfied, neutral, and not satisfied.

Results: A total of 106 patients completed the feedback survey, with 60 in pre-information card phase and 46 in information card phase. Surprisingly, fewer patients in the information card phase were able to recognize at least one physician’s name involving in their care (47.8% vs. 61.7%, p<0.01). 69.6% patients in the information card phase were very satisfied with daily events or plans, compared to 53.3% in pre-information card phase (p=0.04). Furthermore, significantly more patients in the information card phase stated they received more consistent information (95.6% vs. 78.3%, p=0.01). Besides, more patients in information card phase were able to understand the roles of their physicians (43.5% vs. 38.3%, p=0.1), but it was not statistically significant.

Conclusion: Distributing information cards does not improve patient identification of physicians and their roles. However, improved patient satisfaction and physician-patient communication was observed. Alternative economic intervention tools should be considered in the future.

References

Imaging Biomarkers For Improved Lung Cancer Prediction Through Reclassification Of Indeterminate Pulmonary Nodules.

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Introduction: We are facing an epidemic of Indeterminate Pulmonary Nodules (IPNs), with detection of 1.5 million IPNs every year. The probability of malignancy based on clinical and imaging features drives clinical recommendations. Low risk IPNs are followed by imaging and high risk IPNs are referred for tissue evaluation. Management of intermediate risk IPNs imposes a great challenge. Despite the development of a number of predictive models, there still is significant prediction inaccuracy leading to unnecessary biopsies of benign nodules, and potential procedural complications. Here, we tested the hypothesis that semi-automated and quantitative structural imaging analysis allows correct reclassification of IPNs from an intermediate risk group based on the MAYO model to either low (<15%) or high (>80%) risk groups.

Methods: The training cohort of 274 IPNs was developed at VUMC and Nashville VA Medical Center, and the validation cohort of 103 IPNs was independently developed at UPMC. We included IPNs 6-30 mm in longest diameter on less than 2.5 mm thick non-contrast CT scan and with no known history of malignancy at the time of the scan. We compiled clinical data elements for these patients and acquired radiomics features using a semi-automated segmentation radiomics software, developed by HealthMyne. The selected candidate imaging features were summarized as one risk score using LASSO regression model. Bias-corrected clinical net reclassification index (cNRI) was calculated for cases and controls.

Results: 148 cancers and 126 benign nodule were present in training dataset, and 39 cancers and 64 benign nodule controls in validation dataset. The mean largest axial diameter of IPNs for cases in training and validation dataset were 18 mm and 20 mm respectively, whereas for control in training and validation dataset were 12.05 mm and 10.1 mm. AUC for Mayo model on training set was 0.74 (95% CI 0.68-0.80) and on validation dataset was 0.87 (95% CI 0.80-0.94). AUC for combined Mayo and radiomics model, for training dataset was 0.87 (95% CI 0.79-0.95) and for validation dataset was 0.89 (95% CI 0.83-0.95). The biased corrected Net Reclassification Index (cNRI) among cancer was 7.6% in the training cohort and 4.8% in the validation cohort, and for the benign nodules it was 11.1% in the training cohort and 14.2% in the validation cohort.

Conclusion: Our results demonstrate that the integration of clinical and radiomics biomarker in a prediction model allows about 19% reclassification of the IPNs from intermediate risk group to subsequent high-risk or low-risk subgroup. This suggests that structural imaging analysis of IPNs has the potential to improve its non-invasive diagnostic evaluation in specific subgroups.

References

Improving Medication Reconciliation During Hospital Admission

Authors: Dr. Amirtha Dileepan, Dr. Asish Regmi, and Dr. John Pamula

Introduction: Adverse drug events are a leading cause of morbidity and mortality in healthcare systems. Nearly 2/3 of medication lists have one or more errors with resulting costs estimated to be billions of dollars. An accurate, up-to-date medication list is essential to patient safety. Half of errors occur during admission or discharge. We address miscommunication during transitions of care. A standardized process and shared accountability among patient, family, and support staff has shown the best improvements.

We reviewed errors in admission medication reconciliation and the effects of 2 interventions: resident education and a quick "best practice" checklist, and a dedicated pharmacy tech to verify medications with outside pharmacies.

We focused on "high-risk" medications and errors, defined as those involving dual anti-platelet therapy, anticoagulation, insulin, oral hypoglycemics, or opioids.

Methods: We reviewed 6 weeks (42 days) of admissions to the medicine teaching service at Robert Packer Hospital. The on-call resident, nurse, or pharmacy tech collected an updated home medication list which was then reconciled with the medications recorded in EPIC by the admitting physicians. Errors were recorded as addition/duplication, omission, or route/dose/frequency. Results were calculated as total and high-risk errors and errors/admission.

The resident education intervention occured on day 15. The pharmacy tech intervention was on admissions during normal business hours.

Results: Overall there were 108 admissions; 1,215 medications, and 105 high-risk medications. Pre-intervention had 38 admissions, 122 errors in 405 medications (30%), and 11 errors in 44 high-risk medications (25%). Post-intervention had 70 admissions, 136 errors in 810 medications (17%), and 14 errors in 61 high-risk medications (23%). Errors/admission improved from 3.2 to 1.9, and high-risk-errors/admission improved 0.3 to 0.2. We plotted the daily errors/admission for the 42-day study period which showed a 5-day trend with zero errors occuring immediately after the intervention.

63 admissions had no pharmacy tech. These had 71 errors in 586 medications (12%), 5 errors in 41 high-risk medications (12%), average of 1.2 errors/admission and 0.1 high-risk-errors/admission. 45 admissions were with the pharmacy tech. they had 187 errors in 629 medications (30%), 20 errors in 64 high-risk medications (31%), average of 4.2 errors/admission and 0.4 high-risk-errors/admission.

Conclusion: Our study showed resident education led to significant improvement in total errors and non-significant improvement in high-risk errors on admission medication reconciliation. The dynamic data showed the greatest impact was immediately following intervention.

The pharmacy tech was associated with increases in total and high-risk errors. This may reflect improved accuracy of the medication list rather than a true negative impact.

We plan to continue this project by junior residents, which we hope will sustain positive impact and reveal further areas for improvement.
Introduction: Opioid use disorder (OUD) has become a public health crisis in the United States. Prescription opioid pain relievers are among the most commonly misused medicines. Opioids are commonly used for pain management in acute pancreatitis (AP). However, to date, the risk of developing opioid addiction after an episode of AP has not been studied. We sought to determine the incidence of OUD following an episode of AP over an 11-month follow-up period, and its impact on mortality and other outcomes using the largest national readmission database in the United States.

Methods: This is a retrospective cohort study using the newly redesigned 2016 National Readmission Database (NRD). Inclusion criteria were: i) A principal diagnosis of AP and ii) admission in January 2016. Exclusion criteria were age less than 18 years, any elective admission and patients who already had a pre-existing diagnosis of OUD. All readmissions to any hospital for a principal diagnosis of OUD within 11 months of the index admission were recorded for each patient. OUD-related diagnosis were identified using previously published studies. The primary outcome was incidence of OUD. Secondary outcomes were i) OUD-related mortality, ii) OUD-associated healthcare resource utilization [length of stay (LOS) and total hospitalization costs and charges]. Independent risk factors for OUD were also identified using multivariate logistic regression analysis.

Results: A total of 21782 patients were hospitalized with AP in January 2016. The mean age was 51.9 years and 45.8% were females. The 11-month incidence of OUD was 4.4%. After adjusting for potential confounders, the development of OUD following an episode of AP was associated with a significant increase in 11-month all-cause mortality rate [aOR: 2.1, (95% CI: 1.1-4.05) p-value: 0.014]. The total hospital days associated with OUD-related readmissions were 2,466 days, with a total healthcare in-hospital economic burden of $48.7 million. Independent predictors of developing OUD were LOS, patients admitted to hospitals with large volume (fifth quintile) and patients who left against medical advice (AMA). Age, uninsured status and private insurance had a protective effect of developing OUD.

Conclusion: Approximately 4.4% of patients can develop opioid use disorder after an initial episode of acute pancreatitis. The development of opioid use disorder has a substantial impact on mortality and healthcare resource utilization. Our study highlights that opioids should be used judiciously in patients with acute pancreatitis upon discharge from the hospital.
Pre-Encounter Manual Blood Pressure vs Provider Measured Blood Pressure in Primary Care Clinic

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Introduction: Measurement of blood pressure (BP) is fundamental in assessing and treating hypertension, however, methods on hypertension research are often different than clinical practice. Automated office BP (AOBP) was the method used by the SPRINT and other blood pressure trials that were the basis of the 2017 ACC/AHA guidelines on hypertension, which recommends aggressive BP control. Manual office BP (MOBP) measurements are shown to be 7-8mmHg higher on average than AOBP, however, many outpatient clinics continue to use MOBP due to convenience and cost. Patients with BP titrated down with medications to the new ACC/AHA guidelines based on MOBP may be at risk for adverse events. We hypothesized that a repeated manual BP by the physician would be significantly lower than an initial pre-visit manual BP, which may be a more accurate representation of the patient’s true BP.

Methods: In a primary care office, patients were roomed in the usual fashion and a manual BP was measured by nursing staff using an aneroid sphygmomanometer. Initial BPs greater than or equal to 120/80 mmHg were repeated during the clinical encounter by the physician. It was emphasized to nurses and physicians to adhere to ACC/AHA BP measurement guidelines. Exclusion criteria were initial BP less than 120/80 mmHg systolic and age less than 18 years. Comparison between nurse and provider BP were made using the Wilcoxon Signed Rank Test.

Results: Overall, the difference nurse and provider BP were statistically significant with a median difference in systolic BP of 4 mmHg and diastolic BP of 2 mmHg (p<0.0001). The difference between nurse and practitioner measured BP was greatest in patients with initial BPs > 150 mmHg systolic (10 mmHg, p<0.0001), and at least 5 mmHg in all groups with an initial BP ≥ 140 mmHg. There was no significant difference in change in blood pressure based on age. With repeat BP measurement, 34% of patients were able to be reclassified to a lower AHA/ACC hypertension stage with 10% being in a higher stage. Patients initially with stage 1 systolic hypertension were reclassified as controlled (SBP < 130) in 40% of encounters with 11% increasing their hypertension stage. 36% of initial hypertension stage 2 patients were reclassified as stage 1 and 8% were controlled.

Conclusion: In outpatient primary care offices without AOBM, physician measurement of blood pressure following the initial nurse measured BP may provide a lower and more accurate representations of a patient’s true BP. Provider measurement of BP with initial systolic reading greater than 130 mmHg may result in lower stage of HTN. The implications of clinician measured BP in this fashion may be fewer adverse events from overtreatment of BP related to over-aggressive pharmacological BP management approaches and a better reflection of true adherence to standard hypertension performance measures used in the primary care setting.
Adoption of the Coronary Artery Disease – Reporting and Data System (CAD-RADS™) Decreases Downstream Testing and Cardiology Referral in Patients with Coronary Artery Disease Compared To Non-Standardized Reporting

Authors: Joshua M Boster, MD; Robert A Hull, MD; Jeremy M Berger, DO; Michael U Williams, MD; Alec J Sharp, MD; Dustin M Thomas, MD

Introduction: Coronary CT Angiography (CCTA) has emerged as a first line test for stable and acute chest pain, but existing data suggests that CCTA could increase downstream testing, invasive coronary angiography, and subspecialty referral. Angina and ischemia are rare in patients with maximal coronary stenosis < 50%, thus additional testing can be safely deferred in these patients. We examined the impact of a standardized CCTA reporting template (CAD-RADS™), which provides results-based management recommendations, on downstream testing and subspecialty referral.

Methods: CCTAs between 01May2015 and 30Jun2017 were queried (n=2,000), resulting in 1796 CCTAs performed for the evaluation of CAD: 751 using CAD-RADS™ and 1,045 using non-standardized reporting (NSR). Ordering provider specialty, baseline demographics, downstream testing and subspecialty referral were abstracted. CCTAs were interpreted by maximal stenosis: no CAD or stenosis (normal or CAD-RADS 0), 1-49% (nonobstructive or CAD-RADS 1/2), or ≥50% (obstructive or CAD-RADS 3, 4, or 5).

Results: The overall referral rate for additional downstream testing was similar between the groups (10.7% vs 10.8%, p=0.939). Downstream testing was rare in patients with a normal CCTA (3.3% vs 2.2%, p=0.300). Amongst nonobstructive coronary artery disease (CAD) scans in the NSR cohort, downstream testing was more common when compared to CAD-RADS 1 & 2 patients (14.4% vs 5.1%, p<0.001). Overall, appropriate downstream testing was common in patients with obstructive CAD (65.3% vs 49.4%, p=0.052). Referrals for invasive coronary angiography (ICA) were similar between the groups (p=0.638), as was the rate of obstructive CAD on ICA (p=0.193). The incidence of non-diagnostic scans (CAD-RADS N) was higher in the CAD-RADS cohort (9.7% vs 4.2%, p<0.001), potentially resulting in higher downstream testing when compared to NSR patients with a non-diagnostic CCTAs (28.8% vs 11.4%, p=0.038). After excluding normal or non-diagnostic CCTAs, downstream testing was more common in the NSR cohort (23.4% vs 15.5%, p=0.009) without an increase in obstructive CAD detection on ICA (74.2% vs 66.7%, p=0.565). There was no difference in the overall rate of cardiology referral when comparing CAD-RADS to NSR (11.1% vs 10.6%, p=0.817). Cardiology referrals were more common for non-obstructive CAD in the NSR cohort than in the CAD-RADS cohort (14.1% vs 7.5% p=0.012). When excluding patients with normal or non-diagnostic CCTAs, patients in the NSR cohort were more commonly referred for evaluation by a cardiology provider (23.4% vs 15.5%, p=0.009).

Conclusion: Adoption of CAD-RADS™ standardized CCTA reporting template resulted in a reduction in downstream testing and cardiology referral amongst patients with nonobstructive CAD and when excluding normal and non-diagnostic CCTAs without an increase in obstructive CAD detection on ICA.
Texas Research Poster Finalist - Remina Panjwani, DO

Comparative and Predictive Analysis of qSOFA, SOFA, APACHEII and the Number of Ventilator Days in ICU Patients Admitted with Sepsis

Authors: Panjwani, Remina DO; Liu, Luna DO; Walji, Shaista DO; Bohra, Hema DO; Viswanathan, Sahityan MD; Reddy, Prashanth MD; Makhni, Manmeet MD; Balamuthusamy, Saravanan MD; Madhrira, Machaiah MD; Cota, Donna DO

Introduction: Quick Sequential Sepsis-Related Organ Failure Assessment (qSOFA) is a bedside tool to identify patients with suspected infection who are at risk for poor outcome outside the ICU. There is limited data comparing qSOFA and Systemic Inflammatory Response Syndrome (SIRS) scores, however, there is no data comparing SOFA (Sequential Organ Failure Assessment), qSOFA, and APACHE II (Acute Physiology And Chronic Health Evaluation) with correlations to length of ventilator days in ICU patients. The purpose of this study was to analyze each of these scoring systems and their association with ventilator days in ICU patients.

Methods: We performed a single-center retrospective analysis of intubated patients in the ICU from 12/2015 to 12/2017. Patients who met criteria for either SIRS, SEPSIS, Severe SEPSIS, and Septic shock were included in the study. Patients who transitioned to hospice or palliative care were excluded. Linear regression and multiple regression analysis was done comparing SOFA, qSOFA, and APACHE II scores to length of ventilator days, ninety day readmission rates, and ninety day mortality rates.

Results: A total of 241 intubated patients (pts) who met the inclusion criteria were analyzed. Of the 241 patients, 60 pts were readmitted within 90 days and 38 expired within 90 days. Follow up data regarding death out-of-hospital were not available for 53 patients. Pearson's correlation coefficient was used for data analysis which showed poor correlation with SOFA, qSOFA, and APACHE II scores to ventilator days of 0.06 - 0.02, and -0.05 respectively. Correlation of SOFA, qSOFA, and APACHE II scores to 90-day readmission rates were 0.09, -0.02, and 0.04 respectively. Correlation of SOFA, qSOFA, and APACHE II scores to 90-day death rates were 0.32, 0.24, and -0.14 respectively. Multiple regression analysis revealed significant correlations for No. of ventilator days to serum creatinine (< 1.5 and > 1.5), mean arterial pressure (MAP), need for pressors, Hct, altered mental status (AMS) and APACHE 2 scores. Similar correlations were noted with the SOFA scores with creatinine, PaO2, need for pressors and Hct. Based on the MRA, equations have been created that could be used to predict the length of ICU days with ventilator.

Conclusion: Patients diagnosed with SIRS, SEPSIS, Severe SEPSIS and Septic shock and who met criteria underwent SOFA, qSOFA, and APACHE II assessment to prognosticate ventilator days, morbidity, and mortality. Data analysis regarding the correlation of SOFA, qSOFA, and APACHE II scores to ventilator days showed poor correlation in patients admitted to the ICU. Currently, there are no scoring systems available to accurately predict ventilator days, morbidity, or mortality in septic pts. Patients with severe sepsis can decompensate quickly; therefore, further investigation in regards to establishing bedside tools in the ICU is warranted.
TEXAS RESEARCH POSTER FINALIST - DANIEL A HYMAN, MD

Implementing a New Discharge Navigator at the Michael E. DeBakey VA Medical Center

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Introduction: To discharge a patient from the hospital is a complex and multistep process which presents an event point at risk for medical error, but in that, an important opportunity for intervention. Many electronic medical record (EMR) systems have instituted decision support systems in the form of discharge navigators in an attempt to minimize error and increase efficiency of the discharge process. However, CPRS, the EMR in use by the Veterans Administration (VA), does not have a discharge navigator built in. We created a discharge navigator for use within CPRS at the Houston Michael E. DeBakey VA Medical Center (MEDVAMC) with the aim of reducing medication discrepancy rates and increasing the efficiency of the discharge process.

Methods: The discharge navigator was implemented in January 2018 and combined with an educational intervention to train residents on proper use. The rate of discharge medication discrepancy in the year preceding implementation of the navigator was determined by the VA's External Peer Review Program (EPRP) via chart review. Surveys regarding resident satisfaction with the discharge process and perceived error rates were distributed online to inpatient medicine teams. Two weeks after implementation, educational handouts were distributed to reinforce training of the discharge process. A post-intervention survey was distributed and the EPRP discrepancy data was collected until September 2018 and compared to baseline data.

Results: Of 76 total responding residents, 74% used the navigator >50% of the time. The self-perceived rate of discharge mistakes, medication discrepancy, and having to restart the discharge process decreased from 36% to 16%, 30% to 22% and 33% to 23%, respectively. 74% of residents reported that the discharge navigator improved the discharge process. Pre-intervention EPRP showed a 4.2% average medication discrepancy rate, which decreased to 3.2% after intervention. Intervention target of 0% discrepancy rate was achieved by the second month post intervention, however the effect was transient, regressing to the mean after 3 months.

Conclusion: Discharge navigators are a decision support tool that may help to reduce medication discrepancy rates and increase the efficiency of the discharge process. Our data shows that discharging patients at the Houston MEDVAMC is time intensive, has a high rate of perceived initial error and revision, and a measurable rate of final medication discrepancy. The implementation of a discharge navigator was widely perceived to be a process improvement by residents, with self-perceived decreases in medication discrepancy rates. EPRP measured discrepancy rates fell to 0% within two months after implementation, however the effect waned with time, likely due to the rotation of residents present during the intervention to other clinical sites. This problem may be alleviated by introduction of the discharge navigator during intern onboarding. Given the success seen at MEDVAMC, this intervention may be scalable at other VA hospitals.

References

TEXAS RESEARCH POSTER FINALIST - REHAN ANSARI, MD

CAN SMARTPHONES SOLVE THE CONUNDRUM OF POOR HEALTH LITERACY?

Authors: Rehan Ansari MD, Amanda Cantu MD, Laura Garcia MD, Hamzah Shamim, Muhammad Siddiqui, Garry Souffrant MD, James Hanley MD MACP

Introduction: Recently there has been an explosion of smartphone apps with an emphasis to improve healthcare of patients. These apps may significantly improve compliance while enhancing efficiency and value while lowering costs. Our unique population with high levels of poverty and inadequate health literacy are extremely vulnerable to suffer from poor compliance due to suboptimal communication.

Anecdotally we noted many of our patients having smartphones and using them on a daily basis despite a major language barrier. In our initial feasibility study, we hypothesized that in a federally funded clinic patients with inadequate health literacy will have smartphones and would be inclined to use them to improve their understanding and monitoring of their medical conditions.

Methods: This is a cross-sectional study; data was obtained through a survey with randomly selected patients from our federally qualified health center (FQHC) Internal Medicine clinic in Harlingen, Texas. The survey consisted of 12 questions addressing age, reading language, ethnicity, gender, education level, chronic medical conditions, likelihood of app use, types of electronic devices used, app use, types of apps, and health literacy ascertained through the STOFLA method (help reading).

Results: A total of 202 people (24% males and 76% females) participated in our survey conducted in March 2017. 100 identified themselves as English readers and 102 as Spanish readers. The most common age group was 50-65 (41%) followed by 40-49 (27%), 18-29 (12%), 30-39 (10%), and 10% (65+). Education levels had the greatest levels of disparities with 73.5% (34/102) of Spanish speakers having less than a high school degree compared to 23% (18/100) of English speakers. Overall 41% had hypertension, 38% were obese and 38% suffered from diabetes. Smartphone use was similar in both groups, with 80% (80/100) and 78.4% (80/102) use in English and Spanish groups respectively. 81% (81/100) of English readers were using apps compared to only 60% (61/102) in the Spanish group. The most common type of app used was for texts/video calls and communication 66% (133/202) while 44% (82/202) used apps for health. When asked how often someone helps you read hospital/clinic materials, 33.3% (34/102) replied sometimes to always in the Spanish group compared to 18% (18/100) in the English group.

Conclusion: Despite only 26.5% vs. 77% (p<0.01) completing high school and lower health literacy 33.3% vs. 18% (p=0.003), the use of smart phones was nearly identical in our Spanish and English speaking populations 78.4% vs. 80% (p=.204). The FDA recently announced a ‘precertification’ program for mobile apps. Based on our data and previous studies this opens up numerous opportunities for clinicians to use this technology to improve patient care. Further studies will be needed to implement currently available applications and to develop new software addressing health care delivery, literacy and education.

References

**TEXAS RESEARCH POSTER FINALIST - NAVID BERENJI, MD**

**Battle of the Sexes: Cardiac Arrest Edition**

Authors: Navid Berenji, MD, Ashkan Ahmadian-Tehrani, MD, Rushit Kanakia, MD, Ali Seifi, MD

Introduction: The relationship between gender and mortality from cardiac arrest is poorly understood. In this study we compare survivability of cardiac arrest in the United States based on gender.

Methods: We performed a retrospective cohort study on patients with cardiac arrest surveying Healthcare Cost and Utilization Project (HCUP) Hospital Inpatient National Statistics from 2000 to 2014. A total of 262,111 patients with diagnosis of cardiac arrest and ventricular fibrillation (diagnosis defined by ICD-9 sorted by Clinical Classification Software) were surveyed. In-hospital deaths secondary to cardiac arrests were compared between genders. Z-test was used to calculate two-tail p-value to determine statistical difference.

Results: During the study period we reported a total of 262,111 cardiac arrests in years 2000 to 2014. In males, 157,117 of cardiac arrest resulted in 74,050 deaths. In comparison, 104,994 cardiac arrests in females resulted in 61,806 mortalities. Based on these values the combined percentage of mortality from cardiac arrest in years 2000 to 2014 amounted to 47.1% in males as compared to 58.9% in females (p<0.001). Each year within the 2000-2014 demonstrated a higher percentage of mortality by cardiac arrest in females compared to males (p<0.001).

Conclusion: Although the total incidence and mortality by cardiac arrest were shown to be higher in males, the percentages of deaths from cardiac arrest were higher in the female population nationwide from 2000 to 2014. Further studies are warranted to discover the underlying factors that result in the gender difference in survivability from cardiac arrest in order to optimize medical efforts to benefit both populations.
TEXAS RESEARCH POSTER FINALIST - BRIAN DUFFY, MD

Not-so-Smartlinks: Eliminating Superfluous Shortcuts to Reduce Discharge Medication Documentation Discrepancies

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Introduction: Medication errors are prevalent at discharge and contribute to harmful adverse drug events with potentially avoidable healthcare utilization and cost.1-6 Documentation of medications at discharge presents opportunities for several types of errors which may occur in discharge summaries (DS), discharge instructions (DI), or electronically generated discharge medication lists (eDML). Within Epic, our institution’s electronic health record (EHR), each of these document types may contain unique discharge medication lists. While providers could traditionally generate their DS and DI with statically-imported medication lists via Smartlinks, we believe the most accurate medication list is the dynamic eDML. We aimed to identify the frequency of discharge medication documentation errors among these sources and to reduce these errors to zero within one year.

Methods: This Quality Improvement initiative was conducted at Parkland Hospital, a large, tertiary-care county hospital in Dallas, TX. In late 2016, the Hospital Medicine group and Internal Medicine residents were instructed via large group didactics to not import medication lists into their DS and DI. We performed a randomized chart review of 60 discharge encounters from Hospital Medicine and Internal Medicine teaching services between January 1, 2017 and March 30, 2017. We compared medication names, number of medications, dosages, and sigs among the DS, DI, and the eDML for each encounter. After approval by the EHR governance committee, the Smartlinks that import discharge medication lists were disabled in April 2017. We performed a subsequent randomized chart review of 60 discharge encounters between July 1, 2017 and September 30, 2017. Error frequencies before and after disabling of medication list importation were then compared.

Results: Prior to the intervention, 66% of DS contained a medication list, and 15% of DS had discrepancies in medication names, total number of medications, dose, or sig. Post-intervention, these rates were reduced to 35% and 8.3%, respectively. Prior to the intervention, 16.9% of DI contained a medication list, and 6.7% of DI had any of the above discrepancies. Post-intervention, these rates were reduced to 6.7% and 1.7%, respectively. The DS or DI that contained a medication list post-intervention utilized alternative Smartlinks that had not been disabled.

Conclusion: While discrepancies often exist between admission or outpatient medication lists and the discharge medication list,3-6 no studies have described the discrepancies occurring between multiple discharge medication lists generated in the EHR. Here, we demonstrate that multiple EHR sources for discharge medication lists may contribute to error, and that modifying EHR functions may reduce these errors. Further iterations of this project will include disabling additional Smartlinks allowing the importation of medication lists into discharge documentation and ongoing didactics in order to eliminate discharge medication list discrepancies, thereby perhaps improving communication between inpatient providers, outpatient providers, and patients during a high-risk transition of care.

References


Reducing Unnecessary Inpatient Labs in a Teaching Hospital

Authors: Ronak D. Ghiya MD, Harris Lin MD, Trina Dorrah MD, MPH, Michael Ladogana MD

Introduction: In this age of desiring better health care with lower costs, reducing unnecessary laboratory testing can be beneficial to help accomplish both. Unnecessary labs are a substantial contributor to the estimated $910 billion/year in health care waste. Not only is cost an issue but ordering repetitive labs have consequences of further invasive testing that can lead to increased health risks and medical errors for patients. Physicians repeat labs in stable patients because fear of future abnormal labs, laboratory evidence to back clinical suspicion, defensive medicine, monitor the disease process, and clarify thought process to make medical decisions. At our facility the cash pay prices for the following lab tests are: BMP $169, CBC $150, BNP $299, Lipase $94, Amylase $115, CRP $68, and ESR $54.

Methods: Our goal was to reduce unnecessary laboratory test ordering by attending and resident physicians at the Baylor Scott & White Medical Center in Round Rock, TX by increasing awareness of excessive lab ordering during the months of April and May 2018 with reduction of total lab frequency and lab frequency per patient by 10%. For the BMP, CBC, and BNP, the main goal was to reduce repeat ordering. The American College of Physicians (ACP) Choosing Wisely guidelines show that routinely ordering labs daily is not helpful, unless you need them to change management. For amylase/lipase and ESR/CRP, the ACP Choosing Wisely guidelines recommend ordering a lipase instead of amylase and CRP instead of ESR. Lipase and CRP are more sensitive markers for pancreatitis and inflammation, respectively. Our plan for these goals was to increase awareness to attending and resident physicians about being conscious and reducing repetitive daily labs. We placed posters throughout the hospital and discuss this plan with physicians at their monthly meeting.

Results: There was a decrease of approximately 19% in BMP, decrease of 4% in CBC, decrease of 6% in BNP, decrease of 57% in amylase, and increase of 33% in ESR. The benefits of reducing unnecessary inpatient labs are improved patient outcome and satisfaction due to fewer needle sticks and decreased phlebotomy, reduction in medical errors due to abnormal labs, and reduction in costs for patients and the hospital. The potential cost savings from this project within two months was $88,879 from a mere $5 for printing and laminating costs for the posters.

Conclusion: Overall, lab ordering frequency was reduced by 20% and there will be continued education to more health care providers. We plan to incorporate and hopefully reduce other labs (ex: magnesium, troponin, procalcitonin, etc) that are potentially being ordered excessively. In summary, the key learnings are lab testing is expensive, costs can be reduced by ordering daily labs only if necessary, patient safety and satisfaction can be increased, and medical errors can be reduced.

References

Cardiovascular Risk Assessment in United States Subjects for Deployment to Antarctica

Authors: Patricia Rodriguez-Lozano, Mohamed Morsy, Bao Nguyen, Masood Ahmad.

Introduction: Cardiovascular (CV) evaluation has an important role in the medical screening of candidates for deployment in Antarctica under the United States Antarctic Program (USAP). Due to limited medical care and hazards of transportation during harsh winter weather, an adverse cardiac event would endanger subject safety and jeopardize the mission objectives.

Methods: All subjects for deployment are required to pass a physical qualification protocol under the United States Arctic Program (USAP). We reviewed CV history, physical examination, EKG’s, laboratory data, Framingham risk score (FRS), echocardiograms, and stress tests (ST) of all subjects.

Results: From October 2013 to November 2017, 136 subjects were screened for deployment, mean age 48±4.7 years, 24 (17.7%) women & 112 (82.3%) men. Nine subjects (6.6%) had known coronary artery disease. Risk factors included, 24(17.6%) subjects with hypertension, 27(19.8%) with hyperlipidemia, and 44 (32.3%) were smokers. A 10-year risk for CV disease by FRS was calculated. Risk score of < 10 % was considered low, 10% to 20% as intermediate, and > 20% as high. All females had a score of <10%, males had high scores after age 52 years. The risk score was low in 103(75.7%), intermediate in 29(21.3%), and high in 4(3%) subjects. Seventy (51.4%) subjects were recommended to undergo further CV testing. Twenty-five (35.7%) had ST with ischemia in 5(7.1%), 2(2.8%) had normal coronary angiograms, and the remaining tests were unremarkable. Out of a total of 136 subjects, 129 (94.9%) subjects were deployed. None of the deployed subjects had cardiac events during their assignment to USAP stations, follow up period ranging from 8 to 242 days.

Conclusion: Our study, for the first time, describes the application of screening protocols for cardiovascular risk assessment in subjects for deployment to Antarctica. The current screening process for deployment to USAP stations appears adequate in identifying subjects with low risk of cardiac events during the follow up period.
An Evaluation of Critically Ill Patients with Pulmonary Disease Evacuated from Theater Via Critical Care Air Transport Teams (CCATT)

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Introduction: US Air Force Critical Care Air Transport Teams (CCATT) provide advanced medical care during the transport of patients to higher levels of care. The focus of multiple studies has been to evaluate the enroute care provided during CCATT transport of traumatically injured patients. However, almost half of CCATT transports are for non-traumatic medical illnesses. In a previous effort, we reviewed records of patients with a medical diagnosis who were evacuated from theater by CCATT. Pulmonary disease makes up a significant portion of those medical patients who require transport out of theater. Some research exists studying the physiologic effects of altitude and flight; yet, more limited is the information disseminated describing critically ill patients with pulmonary disease transported via CCATT.

Methods: In this sub-analysis, we performed a retrospective review of CCATT medical records for patients who were evacuated from theater due to pulmonary disease and transported via CCATT to Landstuhl Regional Medical Center (LRMC) between January 2007 and April 2015. The data abstracted included demographics, description of current illness, vital signs, labs, in-flight procedures and medications, and in-flight adverse events. Analysis was descriptive to characterize this specific patient population. Overall counts and percentages are reported.

Results: Of the 672 patients who were transported by CCATT with a non-traumatic medical diagnosis, pulmonary was the third most common primary diagnosis (13%) behind cardiac (54%) and neurologic (16%). Of the 84 patients with a primary diagnosis of pulmonary disease, the majority were active duty (67.9%, 57/84), males (84.3%, 70/83) with a median age of 35.5 [24-50] (37.04±13.07) years of age. About 33% (28/84) were diagnosed with pneumonia and 24% (20/84) were diagnosed with a pulmonary embolism. A total 13% (11/84) of patients had a secondary medical diagnosis (Cardiac 55%, n=6; Integumentary 18%, n=2; Renal 9%, n=1; Vascular 9%, n=1; and Immune 9%, n=1). While on CCATT, 50% (42/84) of patients received in-flight ventilator support with the most common mode being AC/SIMV (88%). Of the remaining non-mechanically ventilated patients, 81% (34/42) received supplemental oxygen therapy. For in-flight medications, 13% received paralytics, 48% received analgesics, 51% received sedatives, and 48% received anti-coagulant and/or anti-platelet medications. The most common in-flight unexpected event was respiratory-related (20%), mostly attributable to an increased oxygen requirement (88%).

Conclusion: Patients with pulmonary disease make up the third largest proportion of non-trauma CCATT transports. These patients are critically ill, as determined by the requirement for mechanical ventilation, supplemental oxygen, and increased in-flight oxygen requirement. Further research is needed to guide CCATT training requirements and resource allocation, as well as clinical practice guideline development.
Reducing the inappropriate use of proton pump inhibitors in internal medicine clinic

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Introduction: Proton pump inhibitors (PPI) are one of the most commonly prescribed medications in the primary care setting, and are generally considered to be safe and well tolerated. However, literature has shown that PPI use is associated with a variety of significant adverse outcomes such as pneumonia, *Clostridium difficile* infection, increased fracture risk, drug interactions, and increased cost. The goal of this quality improvement project was to evaluate the incidence and prevalence of PPI prescriptions for non-guideline recommended indications in our residency clinic and to then wean or discontinue their use.

Methods: We performed a review of empaneled internal medicine clinic patients to assess for active PPI use. Using electronic medical records and direct communication with patients, we compiled a list of patients currently using PPIs and the indication for their use. The goal was to determine if the patient had a valid indication for PPI use per ACG and ACP guidelines. Patients without a valid indication were targeted for intervention.

Results: Of a total of 854 patient records reviewed at the initiation of the study, 322 patients were noted to be using PPIs. Of this subset, 217 did not have a recommended indication for their use. Following discussion of the indications for PPI use, the risks and benefits of their use, as well as alternative therapies such as H2 blockers, tapering regimens were initiated.

For moderate to high dose PPI, the dose was decreased by 50% every week until the lowest dose could be achieved, then, if possible, the medication was stopped the following week. At the completion of the 6 month intervention period, 43% of patients were successfully weaned to a reduced dose or no longer using a PPI.

Conclusion: PPIs are widely-used and generally considered to be a well-tolerated therapy for acid-secretion disorders. However, recently concerns have been raised that they are overprescribed and that the possible adverse effects of their long-term use have been under-appreciated. Although patients can be hesitant to discontinue chronic medications even with extensive clinical education, regular discussion and instruction about specific treatments is effective in reducing inappropriate use in the outpatient setting.
A Comparison Study of Internal Medicine Residents and Internal Medicine Hospitalists on Cost-Related Patient Outcomes

Authors: Sean Barnett, D.O., Michael Smith, M.D., Ronald J. Markert, Ph.D.

Introduction: We compared internal medicine resident and hospitalist teams for patients admitted with a variety of common inpatient conditions. We evaluated efficiency and effectiveness of care using the measured outcomes of hospital length of stay (LOS), subspecialty and ICU consults placed, and 30- and 60-day readmissions.

Methods: After IRB approval our retrospective outcome based study was completed at the Dayton VA Medical Center during the six month period from October 2016 to March 2017.

Patients included were all those admitted to either the resident or hospitalist teams for any of the top admitting conditions.

Exclusion criteria were any discrepancy in documentation, abnormal discharge, any other service as primary at any point during admission, and any subsequent admissions during the study period.

Data collected were admit and discharge date, diagnoses, comorbid conditions, subspecialty consults placed, ICU bed utilization, and 30- and 60-day readmissions.

Means and standard deviations are reported for continuous variables, and counts and percent for categorical variables. The independent samples Mann-Whitney Test was used for comparisons involving the two groups (resident vs. hospitalist) and a second variable measured on a continuous scale. The chi square test was used to examine the relationship between two categorical variables (e.g., resident vs. hospitalist and 30-day readmission). Inferences were made at the 0.05 level of significance with no corrections for multiple comparisons.

Results: For our population of 157 patients our analysis demonstrated no significant difference in demographics, rates of readmission, subspecialty consults placed and ICU bed utilization. Our results did demonstrate a statistically significant lower median LOS between the two teams, with hospitalist teams LOS being greater than one full day longer (hospitalist = 5.88±4.06 days vs. resident = 4.72±3.74 days, p=0.019).

Conclusion: The goals of this study were to assess for similarities in efficiency and effectiveness of care between two different internal medicine teams (resident vs. hospitalist) on certain cost-related outcomes. We demonstrated that resident teams treated similar patients and had similar outcomes in all aspects with the notable exception of having a lower median LOS by greater than one full day. This statistically significant difference in hospital LOS could represent a $5000 - $10000 difference in costs to the hospital and potential savings for patients as well. While the results of this single site study should be generalized with caution, it is possible that further evaluation and study in this area could herald a solution to the frightening and increasing physician shortage in the US and the staggeringly high cost of medical care in the US. If further study should substantiate these results it could give strong evidence that an increase in the number of primary care residencies in the US could increase access to care and lower healthcare costs without sacrificing quality of care.

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Fecal Calprotectin Accurately Predicts Histological Remission in Ulcerative Colitis Patients: Potential Role as a Surrogate Biomarker

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Introduction: Mucosal healing is the therapeutic target for ulcerative colitis (UC). However, whether mucosal healing still should be defined by endoscopy, histology, or both is still unclear. Increasing evidence suggests that histological healing may be a better endpoint than endoscopic. Fecal calprotectin (FC) levels may be utilized as a proxy for histological remission. We aimed to assess the depth of UC remission associated with FC levels.

Methods: Retrospective study of UC patients with FC level within 2 weeks prior to colonoscopy. Patient reported outcomes (PRO2), Mayo endoscopic score (0-3), and Nancy score (0-4) were utilized to define clinical, endoscopic, and histological disease activity, respectively. Mann-Whitney U and Kruskal-Wallis tests with receiver operating characteristics (ROC) curve analysis were applied. The association of FC with outcomes of interest were compared using nonparametric tests. The diagnostic performance of FC for EH and MH was analyzed using receiver operating curve (ROC) analysis.

Results: 104 patients were included. 24% had proctitis (E1), 23% left-sided (E2), and 53% pan-colitis (E3). Higher FC levels significantly correlated with Mayo and Nancy scores (p<0.001). Patients with Mayo 0 and Nancy score ≤ 1 (n=22) had significantly lower FC levels compared to those with Mayo 0 and Nancy ≥ 2 (48 vs. 499 p < .0001). FC level of ≤ 50 was significantly associated with histological remission (AUC = 0.91, sensitivity 96%, specificity 50%).

Conclusion: FC levels are significantly correlated with histological activity and reflect microscopic disease activity even in the face of endoscopic healing. A FC level of ≤ 50 µg/g robustly predicted histological remission. Our findings suggest that FC can be used as a sensitive marker for histological healing in UC.
Bleeding Money: Improving the Administration of Anticoagulation in High Risk Patients

Authors: Abhimanyu Chandel, Patrick Bagley, Zachary Gricepatil, Sean Ouimet, Thong Tran, Geoffrey Cole

Introduction: High risk warfarin usage, as defined by a time in therapeutic range (TTR) of less than 65%, has been associated with an increased risk of stroke, systemic embolization, major bleeding, and mortality. Direct acting oral anticoagulants (DOACs) have previously been demonstrated to be both a cost effective alternative and associated with a greater reduction in rates of stroke and systemic embolization during the treatment of non-valvular atrial fibrillation (AF) compared with warfarin. Additionally, DOACs have been associated with a decrease in the rate of major bleeding during the treatment of venous thromboembolism (VTE) and these medications are recommended as first line initial therapy for both indications by major clinical guidelines.

Previous real-world cost data analyses have further suggested that a switch from warfarin to apixaban would result in an estimated total medical cost avoidance of $4440 per patient-year. A recent pilot project demonstrated a safe and effective process of reducing high risk warfarin usage and a similar method was applied to our patient population through this initiative.

Methods: Patients enrolled in the Walter Reed Anticoagulation Clinic who were being treated for non-valvular atrial fibrillation or venous thromboembolism with a TTR less than 65% over the prior year were identified. These patients were screened for a contraindication to DOAC therapy. If no contraindication existed the patient’s primary care physician or primary sub-specialist was contacted with the suggestion that warfarin therapy be discontinued and a DOAC therapy be adopted. The primary outcome evaluated was the change in percentage of high risk warfarin patients enrolled in the Walter Reed Anticoagulation Clinic.

Results: 72 patients were identified as having a TTR less than 65%. 32 of which had no contraindication to DOAC therapy. 31 patients (97%) were subsequently transitioned to a DOAC. Due to this initiative, the total number of high risk warfarin patients was decreased by 42% which resulted in a decrease in the percent of high risk patients enrolled in the Walter Reed Anticoagulation Clinic from 25% to 17%. At three month follow up, all transitioned patients remained on the selected DOAC. No transitioned patients were readmitted for bleeding or experienced thrombotic events. Based on the prior year’s data, as a result of transition to a DOAC, approximately 500 telephone and face-to-face encounters with anticoagulation clinic staff will be avoided per year. Additionally, our intervention was estimated to have resulted in a total annual medical cost avoidance of $136,000 per year.

Conclusion: This project demonstrates an application of a simple, effective, and potentially cost-saving method for reducing the percentage of high risk warfarin within a health system. The impact of projects such as this will likely only increase in the future as the number of accepted contraindications to DOACs decrease and generic alternatives become available.

References


Introduction: Roughly 10% of patients report history of a penicillin [PCN] allergy. However, most of these individuals do not carry a true IgE-mediated allergy to penicillin or its derivatives. Studies have shown higher clinical failure rates, increased hospitalization stay, and increased infection rates of C. diff, MRSA, and VRE for PCN allergy labeled patients. The efficacy and cost-effectiveness of PCN allergy de-labeling is well demonstrated. Current guidelines recommend Pre-pen skin testing, usually performed in an allergy specialist clinic, before oral challenge may be performed. This limits accessibility and promptness of testing, leaving many individuals still labeled as allergic. Our study, conducted from 2017-2018, assesses de-labeling penicillin allergy by skipping directly to oral challenge based off the results of an outpatient questionnaire.

AIM Statement:

1. We are identifying individuals with a documented PCN allergy NMCP internal medicine clinics. We seek to reduce the amount of patients with erroneous or outdated allergies to PCN class antibiotics. Removing this label increases survivability, decreases rates of complicated infections, and decreases the costs associated with increased length of hospitalizations.
2. We seek to de-label 40 patients by July 2018.

Methods: Based off of an in-house questionnaire distributed to Allergy, Internal Medicine, and Infectious Disease clinics, patients are categorized as high, medium, or low risk of true IgE-mediated PCN allergy. High risk patients maintain their allergy status and are not tested. Medium risk patients follow the standardized pre-pen first approach. Low risk patients proceed straight to oral amoxicillin challenge and are observed. All testing performed in the NMCP allergy clinic with full clinic staff and IM epinephrine available for anaphylaxis.

Results: One-hundred and thirty adult questionnaires were collected during our study period. Of these: seventy-one patients were tested in clinic, five questionnaires determined to be high risk, and all others were no shows, cancellations, or refusals. All patients tested were successfully de-labeled with no anaphylactic events.

Conclusion: We have shown that proceeding directly to oral amoxicillin challenge is reasonable in low risk internal medicine patients labeled as having a PCN allergy. Although there were no cases of IgE-mediated anaphylactic reactions in our study, further research is required to verify the safety of our results given the sample size. Further questionnaire alterations may benefit the distinctions of high, medium, and low risk patients.

References

Introduction: Implementation of guidelines have historically led to a significant decrease in mortality, but pneumonia mortality rates have remained relatively constant despite advances in antimicrobial therapy. In 2016 the Infectious Disease Society of America (IDSA) and American Thoracic Society developed new hospital acquired (HAP) and ventilator associated pneumonia (VAP) treatment guidelines. Furthermore, evidence-based order sets have been shown to have the potential to improve pneumonia outcomes (reduced mortality, hospital readmission, and cost).

Methods: The AIM of our quality improvement project was to increase appropriate antibiotic use for HAP, VAP, and community acquired pneumonia (CAP) by 15% comparing a 12 month pre-intervention period to the 12 month post-intervention utilizing the implementation of a standardized admission order set based on current IDSA guidelines. Standardized diagnosis of CAP, HAP, and VAP were as followed: CAP was defined as pneumonia symptoms (cough, sputum production, fever, etc.) plus radiographic evidence; HAP required a 48 hour hospitalization period prior to symptoms; and VAP was defined 48 hours after intubation. Patients were identified by diagnosis code and a retrospective versus prospective cohort was performed comparing the pre (February 2016-February 2017) to post intervention period (April 2017-April 2018). Exclusion criteria included: patients diagnosed at outside facilities, previous treatment initiated, incorrect diagnosis, and immunocompromised patients. “Correct Treatment” was defined as correct medication selection, duration, and dosage.

Results: From February 2016 to February 2017, there were 79 total cases of CAP, 4 cases of HAP and 0 cases of VAP. Pre-intervention, 10 out of 79 cases of CAP (12.66%) were treated correctly and 0 out of 4 cases of HAP were treated correctly. From April 2017 to April 2018, there were 56 cases of CAP, 3 cases of HAP and 0 cases of VAP. 16 out of 56 cases of CAP (28.57%) were treated correctly and 1 out of 3 cases of HAP (33%) were treated correctly resulting in a 15.9% increase in correctly treated HAP and VAP post-intervention, p=0.0123.

Conclusion: In addition to treatment guidelines, the Center for Disease Control has developed “Be Antibiotic Aware” (formerly “Get smart about Antibiotics”) which stresses the importance of starting the right drug, at the right dose for the right duration. Our QI project shows that the implementation of a technological support in the form of an implemented order set to augment provider knowledge improves standard of care toward the ultimate goal of improved patient outcomes in pneumonia.

References

Prophylactic anticoagulation and bleeding risk in ambulatory cancer patients on cancer-directed therapy

Authors: Andrew Wilks MD, Dan Douce MD, Chris Holmes MD PhD

Introduction: Cancer is associated with an elevated risk for venous thromboembolism (VTE), necessitating prophylactic anticoagulation (AC) in cancer patients on cancer-directed therapies. However, AC may lead to an increased risk of clinically significant bleeding events. We investigated whether ambulatory cancer patients on cancer-directed therapies and prophylactic AC had an increased risk of clinically significant major and minor bleeds. Additionally, we investigated if the ATRIA bleeding risk score was predictive of an increased risk of bleeding events in these patients.

Methods: As part of a single-center prospective cohort study enrolling ambulatory patients actively undergoing cancer-directed treatment and being prescribed prophylactic AC, bleeding events and patient demographics were gathered from the medical record. Bleeds and AC dosing were confirmed by physician review. Patients were enrolled from October 2015 through March 2018.

Results: A total of 1,211 patients enrolled in the cohort. The majority of cancers in this population were lung (20.4%), breast (17.6%) and colorectal (13.2%). A total of 82 verified bleeds were recorded, and of these, 44 were major bleeds, 41 were minor bleeds, and 6 subjects had both a major and minor bleed. The odds ratio (OR) of developing a major bleed while on AC was 2.28 (95% CI 1.12, 4.38), while the OR associated with minor bleeding was 0.46 (95% CI 0.32, 2.79). The ATRIA score was not shown to be predictive of major or minor bleeding events, with an AUC of 0.44 (95% CI 0.35, 0.53).

Conclusion: Prophylactic AC in patients on cancer-directed therapy may result in increased risk of major bleeding events. There was no significant increase in minor bleeding events. Additionally, the ATRIA score was not accurate in predicting bleeding events in this cohort. Despite the increased risk of major bleeding events, it may remain advisable to continue prophylactic AC, and these patients should be monitored closely for both VTE and any bleeding event. Additionally, new or modification of existing metrics should be investigated in order to more accurately predict patient on prophylactic AC and cancer-directed therapy who may be at an increased risk of bleeds.
Improving Patients' Ability to Identify Their Physicians Through the Use of Physician Facecards and Whiteboards

Authors: Kramer J. Wahlberg, Shea Lambirth, Zechariah Gardner, Department of Internal Medicine, University of Vermont Medical Center, Burlington, VT, USA

Introduction: More often than not, hospitalized patients are not able to correctly identify members of their physician team. Being able to identify physicians is a critical component of developing the patient-physician relationship. The use of facecards and whiteboards appears to improve hospitalized patients’ ability to identify their physician and may be associated with improvements in patient experience. The aim of this quality improvement study was to assess the impact of implementation of a physician facecard on the ability of patients to correctly identify their attending physician, as well as evaluate current use of patient whiteboards and compare the effectiveness of this visual aid to the facecard. Lastly, we assessed hospitalist perception of these visual aids.

Methods: This prospective study was conducted at the University of Vermont Medical Center. A total of 149 patients and 18 hospitalists were enrolled. Patients were eligible for enrollment if they were admitted to the internal medicine hospitalist teaching service, and hospitalists were enrolled if they were the attending physician on the inpatient service. Hospitalists were randomized to use a physician facecard (intervention) or not (control) during all patient encounters. Patients were surveyed to assess their ability to identify their attending physician and other members of their physician team. Observational data was also collected regarding use of patient whiteboards. Additionally, hospitalists were surveyed to assess their perception of patient’s ability to identify their physicians, as well as attitudes towards use of the facecard.

Results: Patients who received the facecard were significantly more likely to be able to recall the name of their attending physician as compared to the control group (63% vs 32%, p < 0.01). Among the 68 study patients who had the attending physician name recorded on the whiteboard, 68% were able to correctly identify the attending physician as compared to 32% in the control group (p < 0.01). Ninety percent of patients who received a physician facecard and had the attending physician name listed on the whiteboard were able to identify their attending physician correctly, with a regression model demonstrating an independent benefit from each visual aid and no evidence of interaction between them. Eighty percent of hospitalists surveyed agreed that use of the facecard added value and 90% disagreed that routine use of the facecard was burdensome.

Conclusion: The use of physician facecards improves the ability of hospitalized patients to identify their physicians, and the combined use of facecards and whiteboards may provide additive benefits.

References

Perceived Barriers to Guideline-Based Diabetes Care in Hospitalized Adults

Authors: Elizabeth Wahlberg, Allen B. Repp, Department of Medicine, University of Vermont Medical Center, Burlington, VT, USA

Introduction: The prevalence of type 2 diabetes in hospitalized adults is approximately 20%. Treatment of diabetes with scheduled basal-bolus insulin rather than sliding scale insulin regimens is associated with improved glycemic control and reduced length of hospital stay. The American Diabetes Association recommends an insulin regimen with scheduled basal and nutritional doses as the preferred treatment for non-critically ill patients in the hospital. However, utilization of scheduled basal-bolus insulin within the first 48 hours of admission for type 2 diabetics admitted to Internal Medicine Service at UVM Medical Center during the 2016-2017 year was 20%. As part of an initiative to improve inpatient diabetes management, we sought to understand the barriers to guideline-based care as perceived by resident and attending physicians.

Methods: A 26-question anonymous survey was administered to internal medicine resident and attending physicians. Questions were developed based on resident and attending experience as well as review of surveys in the literature. Responses included a Likert-type scale for the frequency that potential factors influence the decision to initiate basal-bolus insulin (“Never” to “Very Frequently”). Statistical analysis was performed using one-way ANOVA to detect differences between groups of training, including interns, upper-level residents, and attending physicians.

Results: Overall survey response rate was 73%. In aggregate, resident and attending physicians reported the most frequent barrier to initiating scheduled insulin was fear of hypoglycemia (mean 3.8/5). Other pertinent barriers included potential changes in diet (3.1/5), expected hospital course too short to start an insulin regimen (2.8/5) and attending preference (2.9/5) or senior resident preference (2.8/5). Interns identified the following barriers as more significant than senior residents or attendings: time for completion of the insulin order-set (p=0.02), preference to defer management to endocrinology (p<0.01) or outpatient care (p=0.02), and uncertainty regarding best practice guidelines (p=0.04).

Conclusion: Similar to prior studies, resident and attending physicians identified fear of hypoglycemia, changes in diet, and short expected hospital course as major barriers to guideline-based insulin ordering. In contrast to prior studies, we found that interns were more likely to identify uncertainty around guidelines and EHR order sets as barriers. Next steps include education tailored to level of training and optimization of the electronic medical record insulin order sets.

References

Fibrocytes in human pulmonary fibrosis: double-blind placebo-controlled crossover pilot study of sirolimus in IPF

Authors: Axell-House DB, Yu V, Zhang Z, Burdick MD, Strieter RM, Mehrad B

Introduction: Idiopathic pulmonary fibrosis (IPF) is a progressive disease associated with poor prognosis. Fibrocytes are a novel population of bone marrow-derived circulating cells that traffic to the lungs and contribute to fibrosis in animal models of pulmonary fibrosis via the interaction of the chemokine CXCL12, made in the lung, and the chemokine receptor CXCR4, expressed on fibrocytes. In human interstitial lung disease, the concentration of CXCR4+ circulating fibrocytes correlates with survival. The expression of fibrocyte CXCR4 is dependent on the mTOR pathway, and is reduced by in vitro treatment with mTOR inhibitor sirolimus. In animal models of lung fibrosis, sirolimus therapy reduced both the trafficking of fibrocytes to the lungs and the extent of lung fibrosis. The effect of sirolimus in human IPF has not been examined. We hypothesized that therapy with the mTOR inhibitor sirolimus reduces the number of circulating fibrocytes in patients with IPF and is associated with an acceptable side-effect profile.

Methods: We performed a short-term randomized double-blinded placebo-controlled crossover study of 30 subjects diagnosed with IPF by consensus definition. Subjects were randomized to placebo or sirolimus for a 1-3 week run-in period with dose adjustment to achieve therapeutic drug levels, then 4 weeks of dose maintenance, and then a 4-week washout period. Subjects then crossed over to the alternate treatment (placebo or sirolimus) for same length run-in, maintenance and washout periods. The concentration of circulating fibrocytes' plasma chemokine ligands, pulmonary function tests, and adverse events were measured at specific time points. Adverse events were classified as serious per NCI CTCAE criteria and were adjudicated as unrelated or possibly related to the study drug by an independent DSMB.

Results: We enrolled 30 subjects between 2011 and 2015; 2 withdrew prior to randomization. Subjects had a median age of 69 (IQR 65-73), 22 were men and 8 were on therapy with pirfenidone or nintedanib. Sirolimus resulted in a statistically significant reduction in circulating fibrocyte concentration (median change -62%, IQR -40% to -29%) whereas placebo did not (median change -32%; IQR -47% to +6%). A total of 49 adverse events occurred during sirolimus treatment and 29 with placebo, of which 35 (2 serious: elevated liver enzymes and angioedema) were adjudicated as possibly related to therapy during sirolimus and 15 (1 serious: worsened dyspnea) during placebo treatments. The incidence of total adverse events, serious adverse events, and total and serious adverse events that were potentially related to therapy did not differ significantly during therapy with drug and placebo.

Conclusion: As compared with placebo, short-term treatment with sirolimus resulted in reduction of circulating fibrocyte concentrations in subjects with IPF and was associated with an acceptable safety profile. Future studies should assess the effect of long-term sirolimus treatment on the natural history of IPF.
Effectiveness of Fecal Immunochemical Testing as the Primary Screening Tool for Colorectal Cancer in an Ambulatory Care Clinic for the Underinsured and Uninsured

Authors: Maurice Marcuard, MD; Edward Oldfield IV, MD; David Mendel; Sami Tahhan, MD; David Johnson, MD

Introduction: The US MultiSociety Task Force (USMSTF) guidelines for colorectal cancer (CRC) screening recommend either colonoscopy or annual fecal immunochemical testing (FIT) as the 1st tier screening methodologies.[1] As a result, the Sentara Ambulatory Care Center (ACC) for the underinsured and uninsured has replaced CT colonography with FIT testing as the preferred screening test. Importantly, the effectiveness of FIT testing is dependent on the quality of the FIT screening program and the ability to follow up positive tests with colonoscopy. To address this, the USMSTF guidelines have outlined several key quality metrics: (1) FIT completion rate to those offered testing of 60% or greater; (2) proportion returning FIT that cannot be processed by the laboratory of less than 5%; and (3) colonoscopy completion rate for those with a positive FIT of 80% or greater.[2] This project was a preliminary analysis of the effectiveness of our FIT testing program at the ACC during the initial year of implementation.

Methods: This retrospective quality improvement project reviewed all ACC patients ordered for FIT testing during the first year of testing (9/1/2016-8/31/2017) and one year of follow-up time (9/1/2017-8/31/2018). Information was obtained by chart review and included all patients ordered for a FIT test without any exclusion criteria. For patients with negative FIT testing, chart review determined if they were able to complete a follow-up FIT test within one year. For patients with positive FIT testing, chart review determined their dates for initial appointment, scheduled and actual colonoscopy dates, and recommended follow up interval.

Results: A total of 308 patients had a FIT test ordered during the study period (26 positive, 230 negatives, and 52 incompletes). The FIT completion rate was 83.1% (256/308). The colonoscopy completion rate for positive FIT tests was 65.4% (17/26), and all patients with a positive FIT had a surgery appointment. For patients with a negative FIT test, only 18% (38/207) of patients completed a follow-up FIT test within one year. Of completed tests, there were 2 FIT tests (0.7%) that could not be processed.

Conclusion

- We found successful adoption of FIT testing for CRC screening with respect to an initial completion rate of 83.1%
- We failed to meet goal rates of colonoscopies getting only 65.4% complete although all patients with a positive FIT test were scheduled an appointment.
- There was a significant drop-off in the FIT testing completion rate after the first year with only 18% of patients undergoing repeat testing
- These results highlight a need to improve colonoscopy completion rates among patients with positive FIT tests and compliance with repeat annual FIT testing for negative FIT tests. To address these deficiencies, we need to explore better development tools for programmatic testing to increase compliance with both outcomes.

References

Comparing Efficacy and Quality of eConsult between Luminal Gastroenterology and Hepatology

Authors: Sa Ra Park, Indira Bhavsar, Jennifer Wang, Anne G. Tuskey, Kimberly Dowdell, Nicholas M. Intagliata, and R. Ann Hays

Introduction: Electronic consultation (eConsult) is an asynchronous, provider-consultant communication platform without consultant-patient contact. It is becoming increasingly popular as it can provide timely access to specialty care. Gastroenterology (GI), both luminal and hepatology, is one of the most frequently consulted specialties through eConsult, but the data regarding types and outcomes of eConsult to GI is limited. In addition, while luminal gastroenterology addresses distinctively different populations and clinical problems from hepatology, previous studies combined these subspecialties together. We aim to evaluate eConsult in these subspecialties separately, and compare the characteristics and outcomes of eConsult between them.

Methods: Retrospective cohort study of all eConsults to GI completed at U.S. tertiary care academic center from January 2015 to May 2017. We reviewed the reasons for eConsult, response time, conversion to a visit with gastroenterologist, and the indirect costs saved through avoided traditional referrals.

Results: We reviewed 304 charts. We excluded 5 charts because 1) eConsult requests did not contain sufficient information to lead to a recommendation, or 2) topics were inappropriate for GI eConsult. Regarding queried topics, luminal GI received inquiries on a diverse range of topics (46 topics in 165 eConsults) and hepatology had a narrow range of topics (5 topics in 129 eConsults). Most commonly queried topics in luminal GI were chronic abdominal pain (26%), colon cancer screening (24%), and chronic diarrhea (18%). In hepatology the most common queried topics were abnormal liver enzymes (55%), abnormal imaging of the liver (23%), and viral hepatitis (18%). Average response time was 2.4 days in luminal GI compared to 1 day in hepatology. Mean wait-time from eConsult placement to face-to-face evaluation was 78 days in luminal and 88 days in hepatology. Hepatology was able to resolve clinical queries more frequently with eConsult alone and avoided more face-to-face evaluation than luminal GI (76% vs 52%). The round-trip mileage (from home to clinic) saved with utilizing eConsult was 50 miles per eConsult for luminal GI and 64 miles per eConsult for hepatology.

Conclusion: Both luminal GI and hepatology eConsultants were able to deliver prompt, high quality, and cost-effective subspecialist care to patients through eConsult. Hepatology was able to resolve clinical queries more frequently than luminal GI with eConsult alone, and this is likely due to of the restricted number of topics that hepatology addressed. For the cases that required clinic visits, eConsultants provided useful recommendations for pre-clinic evaluation which potentially improved the efficacy of clinic visits. Future studies are needed to maximize efficacy and quality of eConsult in both luminal GI and hepatology and study strategies to expand eConsult to community providers.
Glycemia assessed by continuous glucose monitoring and peripheral neuropathy among participants with type 2 diabetes and chronic kidney disease

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Introduction: Glycemic control is risk factor for the development and progression of diabetic kidney disease and diabetic peripheral neuropathy (DPN). HbA1c is a crude marker of glycemia, and continuous glucose monitoring (CGM) may better capture risk of diabetes complications through precisely measured mean glucose, time in target range, and measures of glucose variability. We hypothesized that glycemic dysfunction, measured by CGM, is associated with DPN among participants with type 2 diabetes (T2DM) and moderate-severe CKD.

Methods: We conducted a prospective observational cohort study of 105 people with T2D treated with insulin or sulfonylurea. We enrolled 81 participants with moderate-severe CKD (eGFR <60 mL/min/1.73 m²), and 24 control participants with T2D with eGFR ≥60 mL/min/1.73 m² matched for age, duration of diabetes, HbA1c, and glucose-lowering medications. Each participant wore a CGM for two 6-day periods. DPN was assessed using the Michigan Neuropathy Screening Instrument (MNSI) questionnaire, with a positive MNSI score defined as ≥4 symptoms.

Results: Participants with CKD had a mean age of 69 years, diabetes duration 20 years, eGFR 38 mL/min/1.73 m², and HbA1c 7.7%. The average MNSI score for CKD participants was 3.36, and was 2.33 for controls (difference 1.35 [95% CI 0.03, 2.67], adjusting for age, gender, and race). Forty-four participants reported with DPN (42%) reported an MNSI score ≥4 (26 [69%] with CKD and 18 [41%] controls). Less time in range (70-180 mg/dL by CGM), and higher mean CGM glucose was associated with a higher risk of MNSI score ≥ 4 (OR 1.56 [95% CI 1.04, 2.38] for each additional 331 minutes/day out of range [1 SD], and OR 1.59 [95% CI 1.0, 2.51] for each 40 mg/dL higher CGM glucose [1 SD], adjusting for age, gender, and race, respectively). In contrast, there was no significant association of HbA1c, diabetes duration, or coefficient of variation with DPN.

Conclusion: DPN was common among participants with long-standing T2D and CKD. Less time in range, and a higher mean glucose value were correlated with presence of DPN. On average, participants with CKD had a higher MNSI score than control participants.

Authors: Yousaf Bashir Hadi, MD. G J reynolds, MD.

Introduction: Gastroparesis is a chronic disorder characterized by delayed gastric emptying in the absence of mechanical obstruction. Pylorospasm is thought to be a significant contributor to the disease pathophysiology, and current treatment modalities including botulinum injection, medical therapy, and surgical pyloromyotomy have produced mixed results. In recent years, gastric per-oral endoscopic pyloromyotomy (G-POEM) has gained attention as a possible treatment option, but studies have shown heterogeneous outcomes.

Methods: We conducted a systematic review and meta-analysis of the outcomes of gastric per-oral endoscopic pyloromyotomy. Embase, PubMed, ClinicalTrials.gov, and Cochrane database were searched for published and unpublished studies. All studies that reported outcomes of G-POEM, including case series, retrospective, prospective cohort studies and clinical trials were included. A pooled analysis was conducted for clinical efficacy, scintigraphic efficacy, and safety of the procedure.

Results: Overall, 9 studies including a total of 216 patients have reported outcomes of G-POEM, with a pooled technical success rate of 100%. All studies lacked a control group and were considered before- after self-controlled studies for the purpose of the meta-analysis. Studies reported an average follow up of 8 months (3-24 months).

After intervention with G POEM, patients reported a significant reduction in Gastroparesis Cardinal Symptom Index (GCSI score) at 3 months, (4/9 studies Mean Difference: 1.38, 95% CI [0.26, 2.49]), which was sustained at 6 months, (3/9 studies Mean Difference: 2.04, 95% CI [1.52, 2.56]). Scintigraphic studies showed mean reduction in percentage residual at 4 hr (GES - 4 hr) of 20.52%, (7/9 studies, 95% CI [12.30, 28.74] at follow up.

A total of 24 adverse events were observed; capnoperitoneum was the most commonly observed adverse event (6 cases), and required needle decompression in 2 cases. Bleeding ulcer was reported in 2 patients, and was effectively managed by clip placement.

Conclusion: G-POEM is an effective management option for patients with refractory gastroparesis. It significantly improved subjective symptoms of gastroparesis at 3 months post procedure and the effects were sustained at 6 months after G-POEM. Objective measures of gastric retention also showed a significant improvement with this intervention. Long term follow up data are currently lacking, and randomized controlled trials are needed to establish the efficacy of this management option in comparison to Laparoscopic Pyloromyotomy and GES.

References

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A Comparison of Antibiotic Use Appropriateness Between High and Low Prescribers in the Baltimore Veterans Affairs Medical Center Outpatient Setting

Authors: Brian J. Hopkins, MD; Rohini Dave, PharmD; Jacqueline T. Bork, MD

Introduction: Antimicrobial stewardship (AS) aims to reduce inappropriate antibiotic prescriptions which lead to adverse patient outcomes, increased health care costs, and increased antibiotic resistance. Despite national antibiotic guidelines and efforts made by local AS programs, there continues to be a high prevalence of antimicrobial overuse and misuse in the outpatient setting, with some reports approaching 70%1,2. We aimed to identify factors associated with inappropriate antibiotic use in primary care clinics at the Baltimore Veterans Affairs Medical Center with a focus on the frequency of antibiotic prescribing, an easily captured metric.

Methods: This is a retrospective quality improvement review of outpatient antibiotic orders from October 2016 to September 2017. High (HRx) and low prescribers (LRx) were chosen based on 1 SD above or below the mean of total antibiotic per 1000 encounters, respectively. A sample of antibiotic prescriptions were identified (1:1 HRx:LRx) and reviewed for appropriate indication based on local antibiotic guidelines. Univariate and multivariate logistic regression were performed. Comparisons were made using χ² statistic or Fischer Exact test for categorical variables and Mann-Whitney U test for continuous variables.

Results: There were 1033 total antibiotic prescriptions by 24 prescribers, of which 492 were from the 6 HRx and 152 were from the 6 LRx. Doxycycline (181), fluoroquinolones (135) and amoxicillin-clavulanate (116) were the most commonly prescribed antibiotics. Analysis of 120 sampled prescriptions revealed only 2 statistically significant associations with inappropriate prescription, HRx (OR 2.85, 95% CI 1.35 – 6.04) and cellulitis indication (OR 0.06, 95% CI 0.01-0.27). After adjustment for cellulitis, HRx predicted inappropriate prescription with OR 2.31 (95% CI 1.03-5.21, p=0.0427). When comparing the prescriber groups (HRx vs. LRx), HRx was more likely to have inappropriate prescription by indication (55% vs 30% p=0.0056). HRx more often treated upper respiratory infections (23.3 vs 16.7%, p=0.36) and asymptomatic bacteriuria (6.7 vs 0, p=0.12), though neither was statistically significant. HRx more often prescribed fluoroquinolones (30% vs 6.7%, p=0.001) and antibiotics during winter months (61.7% vs 43.3%, p=0.044). LRx more often prescribed doxycycline (25% vs 1.7%, p=0.0002) and treated cellulitis (23.3% vs. 8.3%, p=0.02). There were no statistically significant differences in the groups otherwise, such as duration of antibiotic, age, sex or race.

Conclusion: HRx in the outpatient setting is associated with an increase in inappropriate prescribing and presents an opportunity for AS intervention. When allocating resources for AS in our institution, surveillance of outpatient antibiotic utilization can help hone in on inappropriate antibiotic prescription. Further investigation is needed to evaluate the best approach in changing prescription behavior in these providers.

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Adult T-cell Leukemia-Lymphoma Presenting as Severe Hypercalcemia
Resident/Fellow Clinical Vignette Podium Presentations
Iatrogenic Vertebral Artery Dissection in a Young Female with Ehlers-Danlos Syndrome

Authors: Faisal S. Ali, MD, Miranda Diana C, MD, Maryam R. Hussain, MD

Introduction: Vertebral artery dissection (VAD) accounts for about two percent of all ischemic strokes in young and middle aged patients. Symptoms of VAD can be subtle, though some patients may present with severe headache and neck pain. Mechanical stretching is a well-established risk factor for dissection of the extracranial segment of the vertebral arteries. This may occur due to hyperextension or rotation of the neck leading to tear in the intima. Patients with connective tissue diseases are at higher risk of vascular intimal tears, and consequently dissection. We report a case of iatrogenic VAD following a dental procedure in a young female with underlying connective tissue disease.

Case Presentation: A 37 year old female with a history of Ehlers Danlos syndrome and chronic migraine headaches presented to the emergency department (ED) due to progressively worsening headache, dizziness and neck muscle spasms that begun three days ago. Her physical examination and vitals were within normal range. Owing to her history, the headache was treated as status migranosus which led to resolution of her headache and she was subsequently discharged to home.

One week later the patient presented again to the ED with persistent neck spasms, constant occipital pain, nausea, phonophobia, lightheadedness, left facial tingling and gait instability. Upon further inquiry it was revealed that her symptomatology was initially triggered while she was undergoing endodontic therapy during which her dentist flexed her neck for a crown placement at which point she felt a sharp tearing pain on the posterior right side of her neck. Following her dental procedure she experienced recurrent headaches and constant neck pain with spasms which were considered recurrent episodes of migraine and were treated as such. A computed tomography angiogram of the head and neck was subsequently performed which revealed an intimal flap, compatible with dissection of the right vertebral artery at C5-C6 level, measuring approximately 1.4 cm. No focal intracranial vascular irregularity or aneurysm was noted. The patient’s neurological symptoms resolved spontaneously after persisting for three hours. Owing to the lack of an aneurysm, no invasive intervention was deemed necessary and the patient was managed conservatively with placement of a cervical soft collar, high dose aspirin, with clinical and serial imaging follow-ups. No further extension of the aneurysm on imaging was observed on 60 day follow-up. She remains hemodynamically stable and without neurological symptoms on 80 day follow-up.

Discussion: VAD is a dreaded pathology which may lead to neurological morbidity and mortality. VAD can develop in patients with connective tissue disease undergoing ipsilateral hyperextension of the neck. A low threshold for further investigation should be kept in such patients who present with subtle symptoms after a potential inciting event, including dental work. Patients who have developed a VAD without an aneurysm can be managed conservatively without surgery and with close follow-up.

References

MICHIGAN CLINICAL VIGNETTE PODIUM PRESENTATION - ANKITA AGGARWAL, MBBS

An unusual cause of diffuse pulmonary hemorrhage

Authors: Ankita Aggarwal, Anubhav Jain, Sarwan Kumar

Introduction: Use of new oral anticoagulants like apixaban has increased since the ARISTOTLE trial revealed that it is superior to the traditional anticoagulant warfarin in preventing stroke or systemic embolism. It was also associated with fewer episodes of major bleeding. Though bleeding episodes remains the most common adverse effect of apixaban, pulmonary hemorrhage has rarely been reported.

Case Presentation: A 75-year-old man with history of chronic obstructive pulmonary disease (COPD) on 3L home oxygen, coronary artery disease and atrial fibrillation presented to the emergency department with complaints of a nonproductive cough, fever, and dyspnea for 1 day. He denied other symptoms including chest pain, orthopnea, paroxysmal nocturnal dyspnea, hemoptysis, pedal edema. On presentation, he was septic with fever (temp: 103), leukopenia (WBC: 3100), tachycardia (heart rate: 100) and hypotension. Examination was unremarkable except reduced air entry with mild wheezing in bilateral lower lobes. Initial labs were normal except leukopenia. Infectious workup including procalcitonin, urinalysis, urine culture, blood culture, sputum culture, influenza A & B test. Chest X-ray showed posterior-inferior lobe infiltrates. CT Chest showed patchy ground-glass in bilateral lower and bilateral posterior upper lobes. IV fluids and empirical antibiotics were started. Patient persisted to have high-grade fever, cough, and dyspnea even after 5 days. Further infectious workup including fungal cultures and antibodies, TB, HIV, legionella, the respiratory viral panel was negative. On day 7, the patient underwent bronchoscopy after holding apixaban. It showed diffuse pulmonary hemorrhage with a suspected source in the right upper lobe. Bronchioalveolar lavage was negative for malignancy, infection. Post-bronchoscopy patient’s apixaban was not resumed. Autoimmune workup including ANA, ANCA, and anti-basement membrane antibody was negative as well. Repeat Chest X-ray 1 month later showed complete resolution.

Discussion: Pulmonary hemorrhage is a rare but life-threatening complication of apixaban. Only 2 cases of diffuse alveolar hemorrhage with apixaban and 4 with rivaroxaban have been reported. Most cases of pulmonary hemorrhage present with hemoptysis. The absence of hemoptysis on presentation made the diagnosis challenging in our case. Due to the absence of any reversal agent, supportive management and sometimes invasive ventilation is the only option. In all the reported cases patients had an underlying pulmonary pathology. In advanced COPD with diffuse emphysema, like in our patient, there is thinning of the alveolocapillary membrane. This may predispose them to hemorrhagic complications. Thus, pulmonary hemorrhage should be considered as an early differential in patients on anticoagulation with underlying pulmonary pathology.
A case of Primary Pulmonary Malignant Melanoma presenting as a Stroke.

Authors: Pramod Jha, MD, Aakanksha Sharma, MD, Abdul Wahab, MD, Ziad Alkhoury, MD, Department of Internal Medicine, Rochester Regional Health/Unity Hospital, Rochester, NY

Introduction: Malignant Melanoma involving the respiratory tract is nearly always metastatic in origin, and primary tumors are extremely rare. Only a handful of cases have been reported to date in the literature. We report a case of a 65-year-old male patient who presented with sudden onset weakness and slurring of speech due to intracranial metastases. He underwent three CT guided biopsies before final diagnosis of Primary Pulmonary Malignant Melanoma was made.

Case Presentation: A 65-year-old man with a 2-month history of a dry cough was admitted with sudden onset of right sided weakness and slurring of speech. Noncontrast Computed tomography (CT) scan of the head showed large intra-parenchymal hemorrhage within the dorsal left frontal lobe and multiple areas of high attenuation throughout the brain parenchyma suggestive of metastatic disease. He had a Chest X-Ray done as an outpatient 5 weeks earlier which was unremarkable. A repeat Chest X-Ray done a week prior to presentation had shown a 5 cm right upper lobe lung mass. CT scan of Chest/Abdomen/Pelvis done on admission showed 5.4 cm right upper lobe lung mass, multiple solid pulmonary nodules in the left lung and a 2.7 cm left adrenal lesion. Three consecutive CT-guided biopsies were performed - as earlier two from right lung mass were non-diagnostic. Third biopsy from one of the left lung nodules was positive for Melanin-A, S-100 and SOX-10 immunostain and negative for Pan-keratin and Synaptophysin suggesting Primary Pulmonary Malignant Melanoma. BRAF V600E mutation was detected. He underwent PET-CT scan which showed hypermetabolic lesions within the brain, lungs, left adrenal and spleen although primary lesion was not identified. He was started on Dexamethasone and Levetiracetam for seizure prophylaxis. He underwent Palliative Whole Brain Radiation Therapy. He was started on Dabrafenib(BRAF inhibitor) and Trametinib(MEK inhibitor) while in the hospital.

Discussion: Malignant Melanoma is a malignant neoplasm of melanocytes, and more than 90% of the reported cases are cutaneous in origin. It has also been described in other mucosal sites and organs, including the oral cavity, paranasal sinuses, esophagus, larynx, vagina, anorectal region and liver. Primary malignant melanoma of the lung is an extremely rare nonepithelial neoplasm that accounts for only 0.01% of all primary lung tumors. Clinical presentation is not different from primary bronchogenic carcinoma. Diagnosis is often difficult and based on set diagnostic criteria. Most patients already have distant metastases at diagnosis and so the prognosis is very poor at best and majority survive less than 18 months.

References

Guillain–Barré Syndrome Occurring After Aortic, Tricuspid and Mitral Valve Surgery: A Case Report and Literature Review.

Authors: Amer Aldamouk, MD, Harneel Saini, MS, and Thomas Marnejon, DO, FACP

Introduction: We present a rare case of Guillain-Barre syndrome (GBS) after aortic, tricuspid and mitral valve surgery and discuss the literature.

Case Presentation: A 53-year-old African American man presented to the ER with acute dyspnea, anxiety, muscle spasms and severe pain in his mid-thoracic spine, along with arm and leg paresthesia. Past surgical and medical history included recent open heart surgery with aortic valve replacement, mitral and tricuspid valve repair and left atrial appendage exclusion 12 days prior to admission. Medical history included ESRD, hemodialysis, HTN, chronic tophaceous gout and hyperlipidemia. Physical exam revealed BP 176/94, pulse 97.9, respiratory rate 20, and BMI 23.89. Cardiovascular exam revealed a well healed sternotomy scar and artificial aortic valve closure on cardiac auscultation. Breath sounds were diminished in the left lung base. Initial CXR revealed elevated left hemidiaphragm. Initial laboratory studies demonstrated Na+ 133, K+ 4.4, CO2 24, Cl 94, BUN 28, Cr 5.9 and blood glucose 93. Complete blood count showed WBC 11.6 with neutrophilia and lymphopenia, Hb 7.0, Hct 20.8 and platelet count 317,000. He subsequently developed symmetric, progressive bilateral weakness and numbness of the lower extremities. Physical exam also revealed areflexia and a sensory level around T 10. CT scan of cervical, thoracic and lumbar spine showed no evidence of compression fracture, hematoma, abscess or transverse myelitis. Lumbar puncture revealed normal opening pressure with clear fluid, protein 315 mg/dL, WBC 5 cells/mm3, RBC 0 cells/mm3, and glucose 62 mg/dL. CSF albumin was 133 mg/dl (0-35) and oligoclonal bands were negative. Acetylcholine receptor antibodies were 0.0 nmol/L. Nasopharyngeal swab for respiratory viruses was negative. Electrodiagnostic studies revealed diffuse sensory-motor peripheral neuropathy with axonal degeneration and demyelinating changes, consistent with GBS. Neurology recommended ICU transfer and IVIG initiation. Stat IgA was normal, 228 mg/dl (70-400). Two days later, respiratory status required intubation and mechanical ventilation. Plasmapheresis was initiated; IVIG was discontinued. Hospital stay was complicated by pneumonia; he was extubated after 7 days. Motor strength gradually improved, he was transferred to acute rehab, and he was discharged home, ultimately regaining full motor strength.

Discussion: CDC GBS surveillance data suggests only a 5% incidence within 8 weeks of surgery. A recent retrospective review of 208 GBS cases demonstrated that 15% of patients developed postsurgical GBS within 8 weeks. In this case series only one patient underwent CABG. Cardiopulmonary bypass has been associated with activation of complement, secretion of both pro- and anti-inflammatory cytokines (IL-8, IL-10), tumor necrosis factor (TNF-α), and activation of neutrophils. Post-surgical GBS incidence may be more common than thought; however, GBS following open heart surgery is exceedingly rare. Clinicians should consider GBS diagnosis in patients who develop post-operative progressive weakness, paresthesia and diminished reflexes.
A Difficult Diagnosis...Acute Mitral Regurgitation Hiding Behind a Community Acquired Pneumonia

Authors: Ali Hasnie MD, Ammar Hasnie MS V, Ragheb Assaly MD

Introduction: Acute Flail Mitral valve leaflet is a time sensitive and reversible etiology of cardiogenic shock. Misdiagnosis is extremely common, with up to 60% of cases being initially misdiagnosed.

Case Presentation: An 81 year old male with past medical history significant for Ischemic Cardiomyopathy, Coronary Artery Disease with stents to the LAD, CHFrEF (EF=30%), DM2, and HTN presented to the hospital with complaints of shortness of breath with associated cough and fatigue. Initial laboratory workup noted leukocytosis of 13,700, Troponin of 0.52, and Procalcitonin of 0.14. Chest X-ray on admission noted some haziness in the right cardiophrenic angle but was otherwise clear. The patient was started on antibiotic therapy for a presumed community acquired pneumonia. While hospitalized he began to complain of increasing shortness of breath and his oxygen requirements began to increase. Repeat chest x-ray noted evidence of developing right middle lobe infiltrate. The patient continued to deteriorate overnight with oxygen saturations of 60% on non-rebreather mask. He was urgently intubated. Antibiotics were escalated, however despite this procalcitonin was noted to have risen to 70 and due to hemodynamic instability he required three vasopressors for adequate perfusion. A trans-thoracic echo performed prior to decompensation noted moderate mitral regurgitation with posteriorly directed flow. Concern arose for Endocarditis with this finding and a trans-esophageal echocardiogram was performed emergently which noted a completely flail anterior mitral valve leaflet and vegetation on the leaflet tip. Cardiac catheterization was performed noting mild to moderate non-obstructive coronary disease of the LAD, LCX, and RCA. Cardiothoracic surgery emergently moved the patient to the operating room for replacement of the flail anterior leaflet with ruptured chordae tendinae with a bioprosthetic valve. Of note, the valve had no evidence of vegetation, and subsequent pathologic evaluation appeared to be consistent with myxomatous degeneration of the valve. The patient was ultimately discharged to home.

Discussion: In this case, the patient’s concomitant pneumonia complicated and delayed diagnosis of his flail anterior mitral valve leaflet. The rising procalcitonin in the setting of increasing vasopressor requirements strongly favored an infectious etiology for the patient’s sudden decompensation. Initial trans-thoracic echo while helpful, was not sufficient for diagnosis. Trans-esophageal echo is the considered the gold standard for the diagnosis of flail mitral valve. Acute flail mitral leaflet resulting in acute mitral regurgitation can result in cardiogenic shock. Prompt diagnosis is critical for the potential for favorable outcome.
Title Multi-organism sepsis following EBUS-TBNA: an unusual complication

Authors: Christine Hsu, Kim Jordan

Introduction: Endobronchial ultrasound with transbronchial needle aspiration (EBUS-TBNA) is a minimally invasive procedure used to obtain tissue from the mediastinal region. Complications are rare; one study of 7000 patients reported adverse events in 1.23% of patients, and bleeding was most common. We present an unusual case of multi-organism sepsis complicating EBUS-TBNA in a young male.

Case Presentation: A 35-year-old male with type 2 diabetes, hypertension, and hyperlipidemia presented with 8 days of persistent epigastric pain, chills, fatigue, and weight loss. Evaluation revealed a pancreatic mass and a peripancreatic lymph node, positive for neuroendocrine tumor. Additionally, right paratracheal lymphadenopathy was seen on chest CT. An EBUS-TBNA was completed and showed scattered lymphocytes, but no metastasis. He was discharged with plans for a Whipple procedure in one month but was readmitted prior to surgery with intractable vomiting and worsened abdominal pain. He also reported new chest pain, chills, night sweats, and myalgias. A repeat chest CT found significant enlargement and necrosis of the right paratracheal lymph node. A second EBUS-TBNA found suppurative inflammation and necrosis. Blood cultures subsequently grew peptostreptococcus species and Preventella denticola and intravenous ampicillin/sulbactam was started. He continued to decline, developed sepsis. Imaging was consistent with a new mediastinal abscess plus new deep vein thrombotic occlusion of right innominate vein and distal right internal jugular. Emergent mediastinoscopy with abscess drainage was performed, and the tissue biopsies now grew Streptococcus viridans, and antibiotics were changed to vancomycin and piperacillin/tazobactam. Infectious disease consultants believed that his septicemia occurred from seeding of oral flora into the right paratracheal lymph node at time of initial EBUS procedure. His hospital course was complicated by empyema, requiring chest tube placement and acute kidney injury, but he eventually recovered from the infection.

Discussion: Infectious complications from EBUS-TBNA are unusual with reported rates of 0.1% to 0.15%. Infection from Streptococcus constellatus, Mycobacterium tuberculosis, Streptococcus pneumoniae, Pseudomonas aeruginosa, Group C streptococcus, Strep viridans, Actinomycyes odontolyticus, Streptococcus mutans, and polymicrobial mixtures with Klebsiella pneumoniae have been reported. In our case, we postulated that direct inoculation of the paratracheal node with oral flora occurred during initial EBUS-TBNA. Risk factors for infection post-EBUS-TBNA are not established but are likely similar to those associated with mediastinitis post-cardiac surgery, including diabetes mellitus, tobacco use and obesity. Though antibiotic prophylaxis and cleaning of nasopharyngeal and oropharyngeal structures by chlorohexidine nasal ointment are routine for prevention of secondary mediastinitis in thoracic surgery, current guidelines recommend against prophylactic antibiotic administration prior to EBUS-TBNA. Physicians must remain vigilant for post-EBUS-TBNA infectious complications, and if occur, choose antibiotics that cover multiple organisms, including those which colonize the oropharyngeal region.
Brilinta Befouls Breathing: Brilinta Intolerance Post Cardiac Cath

Authors: Dr Sharma G (MD), Falahat M (Fourth year MS), Dr Fry J (MD), Western Reserve Health Education, NEOMED, Youngstown Ohio

Introduction: P2Y12 platelet receptor inhibitors such as clopidogrel and (Brilinta) ticagrelor are the cornerstones of treatment of ACS patients [1]. Ticagrelor has been reported to cause dyspnea in more than 13% of patients [2]. We present the case of a 78-year-old female who developed dyspnea related to Ticagrelor in less than 24 hours post cardiac catheterization.

Case Presentation: The patient is a 78-year-old Caucasian female current smoker with past medical history of COPD presented with an acute chest pain. Her pain was centrally located and pressure like. Her initial ECG demonstrated ischemic ST depression and T wave inversion in the anterolateral leads and she was in persistent pain despite narcotics and nitrates. Later that day, she underwent a cardiac catheterization, coronary angiography and balloon angioplasty and a drug-eluting stent implantation. She was then started on losartan, metoprolol, crestor with coenzyme Q10 (given her intolerance to statins) brilinta and aspirin. During her further course of hospitalization her shortness of breath continued to worsen despite her nebulizer treatment, we ruled out all cardiac, pulmonary, metabolic and other causes of shortness of breath. Her Echo was done post catheterization that demonstrated EF of 45%. Her O2 saturation was 99% on 5L oxygen (baseline is room air) via nasal canula and had Cheyne-Stokes breathing pattern. She was afebrile and her CXR was clear, her pro-BNP was not elevated and her Hgb was stable. Brilinta was held on day 3 and she was switched to Plavix. Her shortness of breath resolved, and she was discharged on Plavix with a follow up appointment.

Discussion: Ticagrelor related dyspnea should be suspected in patients who develop shortness of breath soon after starting ticagrelor therapy. Increased sensitivity of peripheral and central chemoreceptor is considered the mechanism of Cheyne-Stokes and dyspnea [1]. It has been reported that dyspnea events were higher in COPD or Asthma patients on Ticagrelor [3]. The higher incidence of dyspnea has been observed with Ticagrelor as compared to other P2Y12 ADP receptor inhibitors is likely explained by differences in pharmacokinetics and reversible binding to receptor (Clopidogrel & Prasugrel bind irreversibly) [4]. Typically, ticagrelor related dyspnea is acute in nature [5]. As was the case in our patient who developed signs of dyspnea in less than 24 hours. In ACS patients on ticagrelor treatment developing dyspnea it is recommended to rule out any other causes of dyspnea and once all causes of dyspnea are ruled out, it is recommended to give the patient some time for possible spontaneous recovery [5]. Only in the case of persistent or severe initial dyspnea should ticagrelor be discontinued [5]. As in previous studies dyspnea associated with Ticagrelor is mostly moderate and transient [5].

References

Fatal Attraction between Giant Cell Arteritis and Acute Coronary Syndrome: is anticardiolipin antibody a cupid?

Authors: Sharmila Bisaria D.O., Khine Shan M.D., Department of Medicine, Hahnemann University Hospital, Drexel University College of Medicine, Philadelphia, PA; Dr. Anubha Tewary, Department of Medicine, Abington Memorial Hospital

Introduction: Giant Cell Arteritis (GCA) is an immune mediated systemic inflammation of medium sized to large arteries, most commonly affecting the aorta and its branches. It is important to understand if GCA serves as a risk factor in patients who develop acute coronary syndrome (ACS) including myocardial infarction (MI) shortly after diagnosis.

Case Presentation: A 76 year old woman with no significant medical history was admitted to the hospital for acute onset of intermittent right sided headache, left sided blurry vision, and transient aphasia. On examination, there were no focal neurological deficits or visual deficits. Initial evaluation for stroke was unremarkable. Angiographic imaging showed arterial wall thickening of the great vessels of head and neck. She was started on prednisone which relieved her headache. Biopsy of the right temporal artery showed transmural granulomatous inflammation consistent with GCA. Within a few hours after biopsy, she complained of chest pain and had anterolateral ST segment elevation on EKG. She was taken for immediate cardiac catheterization; she had no significant stenosis but was found to have apical LAD thrombosis. Her recent ECHO before biopsy was normal. She was started on heparin without stent or angioplasty. Important to note, she had no history of cardiac disease, thrombophilia, or previous clots. However, of 5 gestations she had a history of 1 miscarriage and two stillborn deliveries. She was worked up for thrombophilia and found to have significantly elevated anticardiolipin antibodies (aCL), thus suggesting Antiphospholipid syndrome (APS). She was started on Aspirin, metoprolol and losartan for her ACS and was continued on Prednisone 60 mg daily for GCA. She was transitioned to warfarin for anticoagulation for thrombophilia as use of NOACs in this context is unclear.

Discussion: GCA as a risk factor for ACS is not well understood. Tomasson et al. and Ray et al. have shown an association with increased risk of ACS, peripheral vascular disease, and stroke whereas Ungpraset et al. and Gullo et al. showed no statistically significant increased CAD risk. Manna et al. found increased anticardiolipin antibodies in GCA, although its association with APS is not well established either. The patient’s ACS event post-operatively can possibly be attributed to systemic inflammation or mild thrombophilia or a combination of both. Our case demonstrates a unique presentation of GCA due to its association with APS and ACS. It is important to establish if a correlation exists between APS, GCA, and ACS through exploring more case reports such as this one. Establishing a link between GCA, APS, and ACS may impact therapy including initiation of more aggressive cardiovascular risk modification in those diagnosed with GCA.
Sugar? No, please!

Authors: Kainat Saleem; Haris Zia; Jason Bierenbaum, Department of Internal Medicine, Department of Hematology-Oncology, UPMC McKeesport, PA

Introduction: Immune checkpoint inhibitors (ICIs) such as nivolumab have rapidly become a household name in oncology. Immune-related adverse events (irAE) are a known complication of ICIs. Although severe irAEs are rare (frequency of <1%), they often present as abrupt onset of autoimmune disorders. Now with ICIs being FDA approved for a wide range of malignancies and being used in a rapidly increasing number of patients, a rise in the incidence of these complications is inevitable. We present the first case of an immune-mediated endocrinopathy triggered by escalating the dose of nivolumab.

Case Presentation: A 53-year-old male with no history of diabetes was admitted to the hospital for evaluation of severe fatigue, dizziness, and abrupt weight loss. On admission, he had a random blood glucose level of 536, bicarbonate level of 15, venous pH of 7.30 and beta-hydroxybutyrate level of 5.4. He was diagnosed with diabetic ketoacidosis. Workup revealed a low C-peptide level (0.47), indicative of severe endogenous insulin deficiency. His hemoglobin A1C level was 7.8. Islet cell antibody screen was negative. History revealed that he was being managed for metastatic adenocarcinoma of the lung with nivolumab. He had been taking the fixed-dose 240mg once every two weeks, for one and a half years without any side effects. Two months before presentation, his dose had been increased fixed-dose 480mg once every four weeks. He was diagnosed with new-onset Type 1 Diabetes Mellitus (T1DM) secondary to immune therapy and has maintained on insulin since.

Discussion: With their prevalent use, ICIs are becoming more recognized for their irAE. Nivolumab has recently been FDA-approved for a new, higher (480mg flat every 4 weeks) dose. Literature shows reported cases of new-onset autoimmune diabetes (AD) and fulminant diabetes (FD) in patients treated with nivolumab, with this toxicity generally being irreversible. Pre-treatment HLA genotyping has been proposed to identify patients at risk for FD, however, there are no known markers to identify patients prone to AD. This makes surveillance for AD a challenge and patients usually present with DKA. Based on prior data, the median time of T1DM onset after nivolumab initiation is 2.5 to 4.4 months. Our patient is unique as he tolerated the 240mg nivolumab dose for one and a half year but developed AD within two months of starting the higher dose. This presentation has not been reported in the literature before and opens the question as to whether the higher dose may be associated with an increased risk of irAE, even in patients previously tolerating the lower dose. We advocate for closer monitoring in patients starting treatment with high dose nivolumab, or those transitioning to the higher dose. This should help identify life-threatening complications earlier. Further research is needed to identify patients at risk of these irAEs.

Authors: Abdullah Faisal MD, Mohamed Mortagy MD, Lauren Lamie, DO

Introduction: Bartonella is the most common cause of culture negative endocarditis in the United States which is complicated by kidney failure in 45% of patients. Diagnosing Bartonella endocarditis is very challenging but crucial, because treating Bartonella associated glomerulonephritis (GN) with immunosuppressants is fatal.

Case Presentation: This is a 52-year-old male with a history of multiple comorbidities including a bioprosthetic aortic valve replacement (AVR) who presents to the emergency department with 6 days of bilateral flank pain and hematuria. Physical exam was largely unremarkable including no rashes. Creatinine was 7 mg/dl. Urinalysis was remarkable for gross blood, 100 RBCs, proteinuria and RBCs casts. Imaging of abdomen and pelvis were unremarkable. Autoimmune workup including ANA, ANCA was negative.

Urgent hemodialysis (HD) and corticosteroids were started because of the rapid decline in kidney function. Kidney biopsy was inconclusive due to inadequate tissue. He was discharged on outpatient HD and maintenance steroids taper. One month later, repeat kidney biopsy showed crescentic segmental necrotizing glomerulonephritis and focal deposits of IgM, C19 and C3. He was readmitted for plasmapheresis and a second steroid course and then discharged with maintenance HD.

Over the next 2 months, the patient had multiple episodes of fever, but blood cultures were persistently negative. After four months from initial presentation, he presented to the ED with chest pain and fever. He was diagnosed with NSTEMI, heparin infusion was started and was admitted. Trans-esophageal echocardiogram showed a small mildly calcified echodensity on the bioprosthetic AVR indicating endocarditis and a small echolucent area in the perivalvular region suggestive of an aortic root abscess. The patient subsequently underwent a sternotomy and aortic valve replacement. PCR performed on the vegetation was positive for Bartonella, as well as Bartonella henselae serology was positive for IgG (1/1024) and IgM (> 1/20). He received antibiotics for Bartonella and subsequent serology for bartonella was declining. It is worth mentioning that the patient had a cat which is believed to be the source of the Bartonella infection and he insisted on keeping it.

Discussion: The interval development of endocarditis on echocardiogram, positive Bartonella antibodies and PCR suggest the patient developed glomerulonephritis associated with Bartonella endocarditis. Bartonella is a frequent cause of culture negative endocarditis. Immune deposits showing C3 dominance with IgM staining are consistent with chronic infection related GN.

This case highlights the importance of ruling out infectious etiologies of GN before starting immunosuppressants. As treating an active infection with immunosuppressive agents can be life threatening. We will discuss, infective endocarditis associated rapidly progressive endocarditis, culture negative endocarditis and bartonella endocarditis.

References

Resident/Fellow Clinical Vignette Poster Finalists
Smoke Without Fire: A Case of Thrombocytopenia After Total Knee Replacement

Authors: Seema Kumar, MD¹, Jori May, MD², Rita Paschal, MD²

Introduction - Learning Objectives:

1. Understand the entity of spontaneous HIT and proposed criteria for diagnosis.
2. Recognize orthopedic surgeries as a potential precipitant for spontaneous HIT.

Case Presentation: A 66 year old man with history of hypertension and osteoarthritis presented with three day history of fever, back pain and confusion. He was postoperative day 10 after a bilateral knee replacement. Surgery was uncomplicated and patient was discharged on apixaban prophylaxis. Initial labs were notable for acute kidney injury, hyponatremia, and acute thrombocytopenia with platelet count 50 x10⁹/cmm, decreased from his last recorded value after surgery of 125x10⁹/cmm. His hospital course was complicated by acute respiratory failure and compartment syndrome secondary to extravasation of vasopressors requiring forearm amputation. Doppler ultrasound of the upper extremities revealed nonocclusive thrombi in the left subclavian and axillary veins as well as occlusive clots in left brachial, cephalic and basilic veins. CT imaging showed bilateral adrenal hemorrhage. MRI of the head noted tiny infarcts in the left occipital lobe and left cerebellar hemisphere. Despite management of these acute issues, his platelet count continued to decrease to a nadir of 26 x10⁹/cmm. Although patient had not received any heparin products during surgery or his current hospitalization, his 4T score was 6, suggesting high probability of heparin-induced thrombocytopenia (HIT). Patient was started on bivalirudin for anticoagulation. A platelet factor 4 (PF4)-heparin antigen enzyme linked immunosorbent assay (EIA) showed a strongly positive optical density of 2.9. The serotonin release assay demonstrated heparin-dependent platelet activation and confirmed the diagnosis of HIT. With continued bivalirudin for several weeks, his platelet count improved and no further thrombosis occurred. He was transitioned to warfarin at the time of discharge.

Discussion: Heparin-induced thrombocytopenia (HIT) is a prothrombotic disorder caused by antibodies to complexes of platelet factor 4 (PF4) and heparin. Increasing evidence has established a condition known as spontaneous HIT syndrome, in which HIT develops without prior heparin exposure. Literature review notes a higher frequency of such cases in patients with preceding joint arthroplasty, especially when involving the knee joint. One postulated theory for the pathophysiology involves PF4/heparin antibodies triggered by exposure to knee cartilage glycosaminoglycans, which are structurally similar to the heparin molecule. Proposed diagnostic criteria for spontaneous HIT includes thrombocytopenia (without alternate explanation), thrombosis, lack of proximate heparin exposure, strong positive PF4-dependent EIAs and strong positive platelet activation assay. This patient met all of these criteria. This case highlights the importance of considering HIT in the differential diagnosis of a patient with a consistent clinical picture even without heparin exposure. As the consequences of untreated HIT are life-threatening, appropriate diagnosis and treatment are critical and spontaneous HIT is an important entity to consider.
Vitamin B12 Deficiency Presenting as Pseudo-Thrombotic Microangiopathy

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Introduction: Pseudo-thrombotic microangiopathy (pseudo-TMA) is a recognized, yet uncommon, clinical presentation of vitamin B12 deficiency. Patients with pseudo-TMA present with microangiopathic hemolytic anemia (MAHA), thrombocytopenia and schistocytes. They are often misdiagnosed as thrombotic thrombocytopenia purpura (TTP) and receive unnecessary therapy. Here, we report a case of vitamin B12 deficiency due to pernicious anemia presenting as pseudo-TMA.

Case Presentation: A 60-year-old African American man presented to the emergency room with a two-week history of dyspnea and profound fatigue. Complete blood count showed severe normocytic anemia (hemoglobin (Hb) of 4.2 g/dL, mean corpuscular volume (MCV) was 90.4 µm3, and thrombocytopenia (platelet count of 67,000 cells/mm3). There was also evidence of leukopenia (white blood cell count of 2250 cells/mm3). LDH and indirect bilirubin were elevated and haptoglobin was decreased suggesting ongoing hemolysis. His peripheral blood smear was reviewed and it showed schistocytes and hypersegmented neutrophils. Given the presence of hemolysis, thrombocytopenia and schistocytosis, the diagnosis of TTP was a concern and hence ADAMTS 13 was checked and daily plasmapheresis was initiated. The patient was afebrile and his kidney function was normal. There were no neurologic findings. Given the laboratory findings (pancytopenia, hemolysis and hypersegmented neutrophils) suggestive of vitamin B12 deficiency, a B12 level was checked and it came back low at <60 pg/ml. B12 replacement (1000 mcg IM daily) was started concomitantly with plasmapheresis and parietal cell antibodies and intrinsic factor antibodies were checked. Over the next 3 days, the patient’s hemolysis panel showed gradual improvement. On day 4, ADAMTS 13 results came back normal at 61%. Plasmapheresis was discontinued, and the patient’s clinical status and laboratory findings continued to gradually improve with parenteral B12 replacement. Finally, the parietal cell autoantibodies came back positive consistent with pernicious anemia

Discussion: Patients with severe vitamin B12 deficiency may present with features mimicking TTP such as MAHA, thrombocytopenia and schistocytosis. An early and accurate diagnosis of pseudo-TMA has a critical clinical impact with respect to administering the correct treatment with vitamin B12 replacement and avoiding, or shortening the duration of, unnecessary therapy with plasmapheresis.
As If Peritonitis Were Not Spontaneous Enough: An Underrecognized Complication of Cirrhosis

Authors: Hanjar A1, Fahmawi Y1, Ousley R1, Humphrey K1, Blair J1, Ludvik N1, 1 Internal Medicine Department, University of South Alabama, Mobile, AL.

Introduction: Hepatic hydrothorax occurs in approximately 5-10% of patients with cirrhosis. Spontaneous infection of this pre-existing fluid in the absence of pneumonia is known as spontaneous bacterial empyema (SBE). SBE has been reported to occur in approximately 2% of patients with cirrhosis and in 13-16% of cirrhotic patients with hepatic hydrothorax. This condition is rare, yet has an estimated mortality rate of 30%. Consequently, clinicians need to be aware of this condition so that they can act rapidly to optimize outcomes.

Case Presentation: A 71-year-old Caucasian female with past medical history of cirrhosis secondary to NASH, type 2 diabetes, COPD, and hypothyroidism presented with a one week history of worsening abdominal pain, distention, and shortness of breath. On physical exam, she was afebrile, had diminished breath sounds in the right lower lobe and moderate abdominal distention. Lab evaluation was notable for leukocytosis (11.7 cells/mm3) and lactate of 3.8 mmol/L. MELD-Na score was calculated at 15. Chest radiography revealed a right-sided pleural effusion. CT abdomen-pelvis showed gross ascites. A diagnostic and therapeutic paracentesis was performed. Peritoneal fluid analysis revealed clear yellow fluid with 1,000 RBCs/µL and 176 WBC cells/µL with 7% neutrophils. Peritoneal fluid cultures were negative for any microorganisms. The patient continued to have progressive shortness of breath. A thoracentesis yielded 80cc of cloudy yellow fluid with a pH of 7.55, 43,170 WBCs/µL (88% PMNs), 8,000 RBCs/µL, and a glucose of 83 mg/dL, LDH of 430, protein of 3.5 g/dL. A CT chest with contrast revealed no evidence of mass, pneumonia, or loculation to explain the effusion. Pulmonary service was consulted and the diagnosis of SBE was made. The patient had received meropenem as an inpatient and was discharged on Levofloxacin for a total antibiotic course of 10 days.

Discussion: Spontaneous bacterial empyema remains a frequent yet underdiagnosed condition in patients with hepatic hydrothorax. This is further compounded by SBE occurring in the absence of spontaneous bacterial peritonitis and the limited diagnostic utility of pleural fluid analysis in cirrhotic patients. Prompt diagnosis with thoracentesis for culture and cell count followed by appropriate antibiotic therapy is critical in optimizing outcomes. Antibiotic therapy remains the mainstay of initial therapy for SBE with liver transplantation remaining as the only current definitive therapy. This case highlights the importance of early recognition and treatment in order to optimize patient outcomes.
Vancuishing Cutaneous Small Vessel Vasculitis

Authors: Nabiel Mir, MD; Eduardo Mulanovich, MD; Derek Russel MD; Ryan Kraemer MD

Introduction: Cutaneous Small Vessel Vasculitis (CSVV) is a diffuse palpable, purpuric rash with broad associations. Vancomycin is a commonly used antibiotic, but it is associated with several cutaneous reactions including CSVV. We present a case of Leukocytoclastic vasculitis (LV), a subset of CSVV, which occurred secondary to vancomycin exposure.

Case Presentation: A 55-year-old woman with a history of Sick Sinus Syndrome status post pacemaker insertion presented with a diffuse, pruritic, and painful rash. She was recently discharged from the hospital after being diagnosed with MRSA bacteremia requiring home infusions of vancomycin. Two days afterward, she developed a pruritic, painful rash on her feet that spread centrally over the next 24 hours. She had no mucosal or systemic symptoms. Her physical exam was notable for numerous palpable, non-blanching, circular, purpuric, macular and popular lesions along her extremities and trunk with a few blood-filled bullae. She had a normal CBC, elevated creatinine of 6.0mg/dL, negative urinalysis, and low serum C3. Serologies – including HIV, HCV, HBV, ANA, Anti-dsDNA, Anti-SSA/B, ANCA, Anti-MPO/PR3 – were negative. Echocardiography revealed large tricuspid valve vegetations. A skin biopsy revealed Leukocytoclastic Vasculitis with direct immunofluorescence negative for immunoglobin deposits but positive for fibrin and C3. Her vancomycin was held indefinitely and she was started on daptomycin. The patient's rash resolved over five weeks after the cessation of vancomycin despite multiple readmissions for persistent MRSA bacteremia. Her bacteremia eventually resolved with pacemaker lead removal and prolonged daptomycin infusion over the subsequent 4 months.

Discussion: CSVV typically manifests as palpable purpura involving the lower extremities that can spread to the trunk and upper extremities. CSVV can be caused by chronic infections (e.g. bacterial endocarditis), drug reactions (e.g. vancomycin), autoimmune diseases, and malignancy. In our patient, the suspected culprit was vancomycin because her rash abated with the drug’s cessation while her bacteremia persisted despite the use of alternative antimicrobial agents. The diagnosis for most cutaneous vasculitides is achieved by biopsy of a purpuric lesion. Typical histological findings include an inflammatory infiltrate, vessel destruction, and fibrin deposition. Direct immunofluorescent studies are essential to rule out immune-complex or pauci-immune-mediated vasculitis. LV is a histological subset of CSVV characterized by the presence of neutrophil-predominant infiltrate. Vancomycin-linked LV represents a rare syndrome rooted in hypersensitivity but different from vancomycin’s other cutaneous reactions such as red man syndrome and IgA bullous dermatosis. All reported cases present as a dose-independent, cutaneous-predominant, and centripetal rash that respond to vancomycin cessation. Each case reported variable times to resolution ranging from 24 hours to three weeks. Our case highlights the workup necessary for an adult that presents with a lower extremity purpuric rash and emphasizes the importance of recognizing vancomycin as an uncommon but important cause of cutaneous vasculitis given its widespread use.

References

The Tale of the Insidious Tentacles

Authors: Sarah Tariq, MD

Introduction: Palytoxin is one of the most potent marine toxins known to mankind. It comes from a genus of corals called palythoa zoanthid. MOA: binds the SODIUM/POTASSIUM ATP-ASE pump on the cell membrane disrupting the ion gradient leading to osmotic hemolysis, cytoskeletal actin destabilization, vessel smooth muscle constriction [cardiac]. The rapid influx of extracellular calcium into the cytosol leads to cell damage, followed by death. Exposure is usually via inhalation with boiling of corals, as this aerosolizes the toxin. Symptoms can be vague and affect multiple organs.

**General:** fevers; **Respiratory:** rhinorrhea, cough, bronchospasm, dyspnea; **Skeletal:** myalgia, weakness, tonic contractions, rhabdomyolysis; **Cardiovascular:** bradycardia, ventricular arrhythmias, T-wave elevation, hemolysis; **GI:** nausea, vomiting, **immediate bitter metallic taste;** **Neuro:** dizziness, paresthesia, ataxia, tremors, seizures; **Dermal:** edema, erythema, irritation

Case Presentation: 38 year old male presenting with complaints of sore throat, rhinorrhea, chest pain, and shortness of breath. Onset about 4-5 hours ago; While he was at home doing usual activities; Family had similar symptoms.

PMH: No chronic medical problems. Surgical: NO prior surgeries. Allergies: NKA. Family: NO KNOWN MEDICAL PROBLEMS. SOCIAL: Denies smoking, alcohol or drug use; LIVES AT HOME WITH his 3 sons AND WIFE. Later revealed that he was at home cleaning.

Physical exam was significant for mild distress due to pain, dipahoresis, B/L diffuse wheezing, tachycardia and diffuse muscle tenderness. Labs showed elevated CK and LDH. Chest XR shows no infiltrates of evidence of pulmonary edema.

Discussion: Corals are a gorgeous way to brighten up home aquarium for aquatic enthusiasts. However one thing California law requires Importation permit to import most live aquatic plants and animals, which includes corals. **AZRIZONA LAW STATES THAT IMPORTE WILDLIFE MUST BE lawfully possessed under a valid license, permit, or other form of authorization from another state.**

After involvement of toxicology in our case with suspicion of a toxin as the culprit, home invasion of our patient led to discovery of 100 of corals scattered across the home. The coral was identified and the culprit was isolated as palytoxin.

Treamtent is primarily supportive, including inhaled bronchodilators + steroids to counter respiratory distress, benzodiazepines for seizures and IV fluids for rhabdomyolysis. No antidote currently exists.

The US National Poison Data System revealed 171 cases of inhalation or skin contact with palytoxin between the year of 2000 to 2014.

Obtaining the social history is essential in diagnosis: Inquiring about hobbies, extracurricular activities, work environment. There needs to be more awareness on coral and related toxic exposures amongst medical professionals.
References

Refactory Gastrointestinal Bleed: Old Drug Beats New Technology

Authors: Lavender, C.¹, Lopez, J.¹, ¹: University of Arkansas for Medical Sciences, Department of Internal Medicine

Introduction: Recurrent gastrointestinal (GI) bleeds from arteriovenous malformations (AVMs) are often very challenging to manage. Conventional therapies such as angiographic embolization, endoscopic ablation, surgical resection, and medical management with octreotide or hormonal therapy are often ineffective [1-3]. One randomized trial and multiple case reports have shown promising results for use of thalidomide in refractory GI bleeding from vascular malformations [1,4].

Case Presentation: We present the case of a 67-year-old African-American woman with a significant medical history of type 2 diabetes, hypertension, end-stage renal disease (ESRD), and recurrent diverticulitis who was transferred from an outside hospital for GI bleeding requiring daily transfusions. She reported a three-month history of dark, tarry stools and two weeks of significant fatigue. The physical exam was unremarkable with no abdominal tenderness or distension. Prior to transfer, esophagogastroduodenoscopy (EGD) and colonoscopy were unable to identify the source of bleeding. Once transferred, capsule endoscopy was performed and discovered diffuse, actively bleeding AVMs within the proximal small intestine. Single balloon enteroscopy was subsequently performed and multiple actively bleeding lesions were cauterized with argon plasma coagulation (APC). However, she continued to have melena and decreasing hemoglobin levels over the next week requiring multiple transfusions. Capsule endoscopy was repeated and showed active bleeding from the diffuse small intestinal AVMs. Repeat balloon enteroscopy was performed with another round of APC of AVMs. Despite daily subcutaneous octreotide injections started early in her hospital course, she continued to have episodes of melena and required nearly daily transfusions. Due to the extensive nature of the AVMs, regional small bowel resection was unfeasible. After all of the previous measures failed to halt this patient’s bleeding, a 4-month trial of thalidomide was started. After two weeks of thalidomide therapy, her hemoglobin gradually stabilized and melena resolved. During her third week of therapy, she required no further transfusions (received 19 total over hospital course) and was discharged. At the follow-up outpatient visit two months after discharge, she continued to show no evidence of bleeding, stable hemoglobin, and no adverse effects from thalidomide therapy.

Discussion: This case illustrates the difficult nature of achieving hemostasis in small intestinal AVMs and the benefit of using thalidomide in cases refractory to conventional measures. No specific guidelines exist for GI bleeding due to AVMs refractory to typical endoscopic methods. One randomized control trial with a small sample size, observational studies, and case reports have indicated that thalidomide may be a viable option for refractory GI bleeds from AVMs [1]. Given the excellent response to thalidomide in this case with complete resolution of bleeding when all other attempted measures failed, it is a therapeutic option that should be considered in similar cases.

References

Supplemental Oxygen Usage in an Acute Hospice Care Setting: Blessing or Burden?

Authors: C. Bryan Huang, MD; Roy Kamoga, MD, FACP; Anil Kumar, MD

Introduction: The goal of care for patients admitted to inpatient hospice centers around symptomatic management and comfort care. Medications can provide a provisional level of relief while allowing individuals to spend quality time with family members and loved ones. Continuous oxygen usage in this population may paradoxically prolong life expectancy in some patients who were not hypoxic, potentially raising ethical and moral questions regarding its use in end-of-life care situations.

Case Presentation: A 63-year-old woman with metastatic stage 4 ovarian and colon cancer with obstructive jaundice due to a peripancreatic mass was admitted to inpatient hospice after she did not respond to multiple treatment protocols involving both surgery and chemotherapy. She had uncontrolled generalized pain, intractable vomiting, and profound weight loss. Her vital signs were normal with an examination significant for severe cachexia, jaundice, and lower extremity edema. After admission to inpatient hospice, her life expectancy was anticipated to be no more than a few days. Her initial comfort care regimen included ondansetron, scopolamine, Bisacodyl, a morphine PCA pump with continuous infusion and oxygen at 2L/min. During her stay, her vital signs remained stable despite a progressive functional decline including an inability to tolerating solids and liquids, increasing generalized pain, and a profoundly depressed mental status. It was apparent that she was not at her comfort goal due to a longer than expected dying process while her family sustained continuous psychological and emotional stress. Following request from the family, her nasal cannula oxygen supplementation was removed that resulted in a surprisingly rapid decline in her previously stable vitals, most notably in her oxygen saturation level. The patient expired 1 day later after spending 27 days in inpatient hospice care.

Discussion: The application of supplemental oxygen in palliative care and hospice settings is a controversial topic in patient care on the setting of end-of-life situations. Previous studies have been mixed regarding its use to relieve dyspnea in terminal illness while many patients receive it for “intuition of benefit.” However, one should be cautious over the use of continuous oxygen as it may paradoxically prolong the life expectancy of terminally ill hospice-care patients as demonstrated in this clinical vignette. Subsequent data indicate its usage correlates directly with patient outcome and may act as an unintended, life-sustaining intervention. Such situations can bring about ethical and moral questions whether to continue or discontinue supplementation, while causing undue psychological and emotional burnout on the family, caregiver distress, and an unwanted financial burden. As such, we recommend conservative oxygen use in hospice patients only when associated with discomfort due to dyspnea in comfort care settings.

References

Getting High on Loperamide

Authors: Manojna Konda, M.D., Gayathri Krishnan, M.D., Kulsum Bano, M.D., Richa Parikh, M.D. and Katie Defore, M.D., Department of Medicine, University of Arkansas for Medical Sciences

Introduction: Loperamide is an over-the-counter inexpensive antidiarrheal medication that is a potent intestinal opioid receptor agonist. It was thought to have little misuse potential as it does not readily cross the blood-brain barrier when taken at recommended dosing of maximum 16 mg/day. However, when taken in large amounts, it can cross the blood-brain barrier and cause euphoria or “high”. We present an atypical case of acute opioid withdrawal caused by naltrexone, an opioid antagonist, in a patient who was abusing loperamide.

Case Presentation: A 73-year-old man with history of opioid abuse was transferred to our facility from an outpatient detox center for seizure-like activity and explosive diarrhea after he took naltrexone. He denied any recent opioid use. His urine drug screen obtained before he was started on naltrexone was negative for opioids or any other drugs. Shortly after he took his first dose of naltrexone, he developed generalized shivering of his body lasting about 10 seconds with no loss of consciousness. He also complained of abdominal cramps with vomiting and several episodes of diarrhea. His work-up including complete blood count, basic metabolic panel, liver function test, ECG were normal. The patient later confessed to taking about 80 pills (160 mg) of loperamide a day to “get high” for the past month after he found about it on the internet. The patient’s presentation was consistent with acute opioid withdrawal precipitated by naltrexone. He was admitted overnight for monitoring due to association of loperamide with arrhythmias. He continued to have several episodes of diarrhea and was later discharged to an outpatient substance abuse program.

Discussion: As prescription opioids have become less accessible, the use of loperamide as an opioid alternative is on the rise as it is inexpensive and readily available over-the-counter. It is relatively safe when taken at the recommended dosing for acute or chronic diarrhea. Recently, there has been increasing discussions on the internet regarding using high doses of loperamide to self-treat opioid withdrawal symptoms or to attain feelings of euphoria leading to its increase in popularity in substance abusers. However it is not widely known that when taken at higher doses, it can lead to QT prolongation, life threatening ventricular arrhythmias and cardiac arrest. Unfortunately, currently there is not much information about treating loperamide toxicity. Furthermore, loperamide is not detected in the routine urine drug screen for opioids and requires special serum testing. It is also important to note that there is a possibility of withdrawal which can be provoked by opioid antagonists. Health care professionals need to be aware of the potential for loperamide misuse. A thorough history of use of over-the-counter medications should be obtained in patients with potential for substance abuse.
CURIOUS CASE OF ALTERED MENTAL STATUS - NOCARDIOSIS IN A PATIENT WITH ECTOPIC ACTH SYNDROME

Authors: Krishnan, Gayathri; Ananthulla, Aneesha; Wood, Cole; Goraya, Harmeen

Introduction: Nocardiosis in the context of ectopic adrenocorticotrophic hormone (ACTH) is very rare and to the best of our knowledge, only 12 cases have been reported so far. Here we present a case where the diagnosis was complicated by various manifestations of hypercortisolism.

Case Presentation: A 50 year old Caucasian female with depression, peptic ulcer disease, prior partial gastrectomy, recently diagnosed diabetes mellitus, refractory hypertension, osteomalacia and multiple pathological fractures was referred to our facility due to altered mental status. In addition to above, she had noticed about 50 pounds weight gain over 3 months, easy bruising and amenorrhea for 6 months. Her mental status progressively worsened after admission and CT brain showed multiple ring enhancing lesions. Lumbar puncture showed 1 WBC, increased protein, normal glucose and negative bacterial, fungal and AFB cultures. Meningitis PCR panel, multiple sclerosis panel, Cryptococcal antigen and Toxoplasma antibodies were negative. She was initially started on vancomycin, ceftriaxone, fluconazole and metronidazole. Transesophageal echocardiography was negative for vegetations. CT chest, abdomen and pelvis showed infiltrates in the lung, multiple bilateral non-calcified nodules, thickening of bilateral adrenal glands and a pancreatic lesion. Her cortisol level after 1mg dexamethasone suppression test was >123 mcg/dl and ACTH was 658 pg/ml. ACTH remained elevated even after high dose dexamethasone suppression test. MRI pituitary protocol did not show pituitary adenoma. She was deemed high risk for bilateral adrenalectomy and hence, started on ketoconazole and metyrapone for ectopic ACTH syndrome, but the source was never identified. Subsequently, she developed respiratory failure and had to be intubated. Bronchoscopy with BAL culture grew gram positive branching filamentous bacteria suggestive of Nocardia. Fungal blood cultures by this time resulted Nocardia farcinia. She was diagnosed with nocardiosis with dissemination to the brain. She was ultimately treated with IV trimethoprim sulfamethoxazole, IV moxifloxacin and p.o linezolid. She was continued on these antibiotics but developed multiorgan failure. Ultimately family pursued comfort care measures and she expired.

Discussion: Ectopic ACTH production can lead to high levels of corticosteroids which can suppress host cell mediated immunity and result in hampered host defense against infectious pathogens. Nocardiosis has been described in patients with AIDS, solid organ or bone marrow transplant recipients, patients on chemotherapy and long term corticosteroids and to a lesser frequency in Cushing’s syndrome and immunocompetent patients. Nocardia farcinia is a weakly gram positive, partially acid fast aerobic actinomycete which can cause pulmonary and disseminated infections. This case was interesting since she had a new diagnosis of Cushing’s syndrome, was immunocompromised from endogenous corticosteroids and her presentation was complicated with concurrent infection. It is important to recognize that patients with Cushing’s syndrome are prone to opportunistic infections and brings to light the need for early treatment and prophylaxis.
Hemophagocytic Lymphohistiocytosis: A Case of Fulminant Hepatic Failure in a 50-year-old

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Introduction: We describe a case of hemophagocytic lymphohistiocytosis (HLH) in a 50-year-old man evaluated in the St. John’s, Newfoundland, Canada for fever, sudden onset hypotension, hypoglycemia, jaundice and decreased level of consciousness.

Case Presentation: The patient was enroute to another tertiary care centre for further workup of fulminant liver failure of unknown etiology and a possible liver transplant when he clinically deteriorated. He was an otherwise healthy individual with a history of a possible viral URTI preceding his symptom onset.

His course in ICU was complicated by pancytopenias, acute kidney injury with profound acidosis, recurrent GI bleeds and DIC. Stabilization required intubation and ventilation, broad spectrum antibiotics, multiple vasopressors, blood product transfusions and continuous renal replacement therapy. Nephrology, hematology and gastroenterology consult services were involved in the patient’s care. Despite appropriate supportive measures for two to three days, the patient’s fulminant liver failure and DIC remained persistent.

Repeat comprehensive workup for fulminant liver failure was negative including a viral hepatitis panel, autoimmune serology and an absence of an acetaminophen/ischemic hepatitis history. Abdominal CT imaging showed hepatic steatosis with no obstruction of biliary/hepatic ducts or portal vein. Our comprehensive workup yielded a Ferritin level of >1,000,000, raising the suspicion for HLH.

In retrospect, the patient met 5 out of 8 criteria for HLH, namely fever, pancytopenia, low fibrinogen, elevated ferritin and hemophagocytosis on marrow biopsy. Other criteria not fulfilled were splenomegaly (history of splenectomy secondary to spherocytosis), triglycerides (serum was too icteric), NK function (assay not available) and CD25 levels (referred to outside laboratory).

A Canadian HLH expert was consulted and suggested steroids with consideration for chemotherapy with clinical improvement. Following good response to high dose steroids, the patient’s family consented to treatment with Dexamethasone and Etoposide. With stabilization and signs of improvement with treatment, the patient was transported to another facility for further treatment and consideration of a liver transplant. Despite continued treatment for HLH, the patient passed in hospital due to fulminant liver failure. Autopsy was declined by the family.

Discussion: In our discussion, we review the differential for fulminant liver failure and extremely elevated ferritin. We will also provide a brief overview of our current understanding of the pathophysiology of HLH alongside the diagnostic and treatment standards.
‘Case to Surprise - Lady presented with expressive dysphasia’

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Introduction: Expressive dysphasia is one of the commonest presentations of acute stroke occurring one third presentations of stroke in the world [1]. This is an interesting case of progressive dysphasia in an elderly lady who was eventually diagnosed as a case Creutzfeldt–Jakob disease- (CJD), a fatal neurodegenerative condition, caused by Prion protein. The aim of this case report is to highlight expressive dysphasia as 1st presentation of CJD, initially mimicking stroke and thereafter, rapid evolution of symptoms and positive investigation results lead on to the diagnosis of Creutzfeldt–Jakob disease.

Case Presentation: A 69-year-old lady presented with sudden onset expressive dysphasia, initially with stuttering and anomic aphasia. With a background of multiple vascular risk factors, she was clinically diagnosed as Stroke (PACS). After 2 weeks, she was again admitted with aggravation of dysphasia and new onset confusion. She was very slow in response with further deterioration of speech including both receptive and expressive dysphasia. In this admission, she was found to have myoclonic jerks predominantly in the right arm and asymmetric lead pipe rigidity. Gradually she developed palmo-mental reflex, finger apraxia and echolalia. The gait remained normal and there was no limb weakness and reflexes were intact. Over a period of a week, there was impairment in vigilance, significant memory loss as unable to recognize family members and developed an abulic-akinetic mutism. Routine bloods were unremarkable. MRI of brain revealed DWI and T2 asymmetrical cortical thickening in occipito-parieto-temporal region and hyperintesity of left caudate nucleus. EEG revealed excess of semi-periodic epileptiform discharges over the left hemisphere, slow wave activity widespread. Clinical and radiological suspicion of sporadic Creutzfeldt-Jakob disease (CJD) was confirmed with a positive CSF14-3-3 and CSF RT-QuIC done in collaboration with Edinburgh CJD research team.

Discussion: Creutzfeldt-Jakob disease belongs to a group of prion diseases that leads to development of a panel of complex neurological changes in the cortex. This case is reported as aphasia being a very rare 1st presentation of CJD. [2] Many researches are being carried out to bring about a revolutionary treatment but yet to be effective to improve survival. New Hopes in the Horizon with PRN 100, an artificially manufactured antibody developed by Prion Unit at University College London (UCL) , designed to bind tightly to normal proteins in the brain aiming to prevent abnormal prions from being able to attach themselves to healthy proteins, preventing degeneration of brain. [3].

References

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That “MAHA!” moment: Recognizing a rare presentation of cobalamin deficiency.

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Introduction: Cobalamin (vitamin B12) deficiency can be caused by insufficient intake, decreased absorption as seen after ileal and bariatric surgery, iatrogenic effect of medications, and by autoimmune conditions such as pernicious anemia. This can lead to well-known problems such as macrocytic anemia, pancytopenia, and subacute combined degeneration. More rarely it can cause a pseudo-thrombotic microangiopathy, which presents with microangiopathic hemolytic anemia (MAHA), thrombocytopenia, fevers, and organ dysfunction.

Case Presentation: A 53-year-old male presented to his primary care doctor with weight loss, weakness, and bloody stools for two weeks. His initial outpatient work-up showed anemia with a hemoglobin concentration of 5.4 g/dL and thrombocytopenia to 117,000 µL. He then presented to the emergency room and was admitted for further work-up. His vital signs revealed he was afebrile, with a blood pressure of 126/61 and a heart rate of 84. A rectal examination did reveal brown stool that was guaiac positive. He was immediately transfused with two units of packed red blood cells with improvement in his hemoglobin concentration to 7.2 g/dL. MAHA was suggested by a significantly elevated lactate dehydrogenase (LDH) at 6720 U/L, an elevated d-dimer to 3.07 mcg/mL and the presence of moderate schistocytes on peripheral smear. Although thrombotic thrombocytopenic purpura (TTP) was considered, the presence of macrocytosis and rare hypersegmented neutrophils suggested an alternative diagnosis: pseudo-thrombotic microangiopathy due to cobalamin deficiency. Further work-up revealed a normal folate level, however the cobalamin level being significantly low at <150 pg/mL, with high levels of both homocysteine at 98.3 µmol/L and methylmalonic acid at 53,940 nmol/L. His anti-parietal cell and intrinsic factor antibodies were high and normal respectively. To assist in differentiating the two differential diagnoses, we checked the reticulocyte count which was 1.37% and corrected reticulocyte count of 0.43%. Given suspicion for gastrointestinal bleeding, he underwent an esophagogastroduodenoscopy (EGD) which revealed mild chronic gastritis without significant bleeding. A colonoscopy was also performed and was non-diagnostic. He was started on daily cobalamin injections for five days, after which he was discharged and prescribed monthly injections. Within 11 days post-discharge, his platelet count had normalized while his anemia resolved after two months.

Discussion: This case highlights pseudo-thrombotic microangiopathy as a rare complication of pernicious anemia. Although this entity and TTP present similarly, case reports suggest that reticulocytopenia strongly suggests microangiopathy due to cobalamin deficiency. Because the high mortality rate associated with untreated TTP prompts early intervention, it is important to recognize pseudo-thrombotic microangiopathy due to cobalamin deficiency to avoid unnecessary initiation of plasmapheresis or steroids.

References

A Unique Case of Hyperviscosity Syndrome

Authors: Karen Haiber, MD, Matthew Johnson, MD, and Patrick Sarte, MD, MS, FAAP, FACP

Introduction: Hyperviscosity syndrome (HVS) is a rare clinical syndrome characterized by neurologic abnormalities, vision changes and mucosal bleeding due to high blood viscosity. Typical causes include plasma cell dyscrasias, leukemia and myeloproliferative disorders.

Case Presentation: A 57-year-old male with a history of chronic Hepatitis C presents with one month of worsening weakness, dizziness, gum bleeding, and 10-pound unintentional weight loss. Lab findings were significant for a leukocytosis with lymphocytic predominance and an elevated protein gap. The clinical picture was concerning for hyperviscosity syndrome (HVS) secondary to an undiagnosed hypergammaglobulinemia. The patient received plasmapheresis with improvement in his symptoms. Work-up demonstrated a monoclonal IgM paraproteinemia and an elevated serum viscosity. Staging PET CT showed diffusely increased metabolic activity in the bone marrow without evidence of metastatic disease. Bone marrow biopsy was preliminarily consistent with a low-grade B-cell lymphoma with plasmacytic differentiation. At that time, genetic testing was pending, which would differentiate between a Marginal Cell Lymphoma and Waldenstrom's Macroglobulinemia (WM). Given that the initial treatment regimen is the same for both diseases, the patient was started on Bendamustine+Rituximab+Dexamethasone prior to a final pathologic diagnosis.

Discussion: The most likely diagnosis in our patient was Waldenstrom’s Macroglobulinemia, a lymphoplasmacytic lymphoma in the bone marrow with an IgM monoclonal gammopathy in the blood that can present with HVS due to markedly elevated IgM. The exclusive bone marrow involvement is also typical of WM. However, the final bone marrow biopsy showed low-grade B-cell lymphoma consistent with splenic marginal zone lymphoma (SMZL). Genetic testing showed a lack of an MYD88 mutation which suggested against a diagnosis of WM. Distinguishing marginal zone lymphomas from other small B-cell lymphomas with plasmacytic differentiation, especially WM, can be challenging due to the lack of specific morphologic, immunophenotypic, or chromosomal markers. Both SMZL and WM can present with an IgM paraproteinemia; however, the levels of IgM in SMZL usually remain low, and HVS is very unusual. On the other hand, up to 30% of patients with WM can develop HVS. Despite pathologic and genetic confirmation of SMZL, this case highlights a clinical presentation more consistent with WM. Typically, SMZL presents with prominent extramedullary involvement while WM exclusively affects the bone marrow, which was seen in our patient. The magnitude of monoclonal IgM production and serum hyperviscosity in our patient is also atypical for SMZL. Distinguishing the diagnosis is important, given the majority of SMZL will have an indolent course with a median survival of 10 years and future treatment options may include splenectomy.
A Case of Statin Induced Hyperglycemia

Authors: John Webster, DO, Eugene Han, DO, Brandon Chock, MD

Introduction: Statins can lead to worsening glycemic control and the development of diabetes. While large meta analysis studies show an increase risk in diabetes incidence with statin use, it is important to note that other trials have shown that the benefits of statins offset the risks of diabetes when measuring reduction in myocardial infarction, stroke, admission to hospital for unstable angina, arterial revascularization, or cardiovascular death.

Case Presentation: A 68 year old man with past medical history of hyperlipidemia, hypertension, hypothyroidism and diabetes contacted his Primary Care Physician (PCP) with polyuria and hyperglycemia into the 500s. Of note, he had first developed diabetes in 2008 after a steroid taper and by 2013 had stopped insulin and continued only metformin monotherapy. He had also been on statins for many years. In 2015 he decided to stop both his statin and metformin and his A1C remained stable off medication (A1C 6.8-7.2). He had made no changes to his diet or exercise routine. In response to the patient’s polyuria, his PCP checked an A1C which came back at 11.7. His PCP started him on metformin/glipizide. The PCP soon remembered that he had started the patient on Lipitor several months prior and decided to stop the statin therapy as well. Within a month the A1C decreased to 7.2 and both metformin and glipizide were soon discontinued.

Discussion: While the mechanism by which statins increase diabetes risk remains unclear, it has been shown that patients with diabetes risk factors are most affected. Studies looking at the impact of statins on glucose metabolism are conflicting. A link has been established between the reduction of HMG CoA reductase activity and the incidence of diabetes thus there is evidence that the statin mechanism of action may be related to this increased diabetes risk. Furthermore it has been shown that high dose statins confer an increased risk compared to moderate intensity statins which further supports this notion. Other factors include genetic polymorphisms, length of time on a statin and patient behavior regarding diet and exercise.

Ultimately, statins are recommended to diabetic patients because their cardiovascular benefits far outweigh the negative impact on glucose control. However this case underscores the importance of monitoring for worsening hyperglycemia while a patient is on a statin. Interventions to be considered in these cases include diet and exercise counseling, trying a lower potency of statin or lower dose of statin or trying one of the statins less associated with hyperglycemia.
Difficult to swallow: a unique presentation of IgG4 Related Disease

Authors: Minh-da Le, Megan Cochran, Vera Vaninskaya, Vineet Gupta, Brian Kwan

Introduction: Immunoglobulin G4 related disease (IgG4-RD) is a systemic fibroinflammatory condition characterized by positive lymphoplasmacytic infiltrations involving single or multiple organs.

Case Presentation: A 44 year old man with active tobacco and alcohol use presented with progressively worsening dysphagia to both solids and liquids and a 100-pound weight loss over six months. Physical examination was significant for scaphoid abdomen. Initial labs were notable for hypernatremia 147, hypokalemia 3.0, and acute kidney injury (BUN 63, creatinine 2.97). CT of the chest, abdomen, and pelvis demonstrated mild distal esophageal wall thickening that tapered to a close at the gastroesophageal junction and non-specific prominent mediastinal lymph nodes. Esophagogastroduodenoscopy revealed a high grade mid/distal stricture that was histologically consistent with a benign etiology. IgG4 immunostain demonstrated “focally more than 70 IgG4 positive plasma cells”, suggesting a diagnosis of IgG4-RD. A serum immunoglobulin panel was within normal limits with IgA 141 (RR 70-400), IgG 1394 (RR 700-1600), and IgM 125 (RR 20-230), but IgG4 was elevated at 141 (1-123). The patient was started on high dose intravenous solumedrol (32mg daily) and received an infusion of Rituximab (1g). A gastrostomy tube was placed for severe malnutrition. The patient was discharged on high dose prednisone (60mg daily) with plans for a second Rituximab infusion. His stricture persisted and required outpatient esophageal dilations.

Discussion: Although IgG4-RD was first described in the context of type 1 autoimmune pancreatitis, it is now widely accepted that IgG4-RD can affect nearly any organ system. Characteristic manifestations include sclerosing cholangitis, retroperitoneal fibrosis, sialadenitis and aortitis. Esophageal involvement is very rare. This case highlights several important features. First, IgG4-RD typically has a subacute presentation mimicking a mass lesion of the affected organ. Second, histopathology has a central role in the diagnosis of IgG4-RD. While serum IgG4 levels can be elevated, they are non-specific and are seen in other autoimmune disorders. Without the use of appropriate immunohistochemical stains, the diagnosis of IgG4-RD esophagitis could have been easily missed. Third, early initiation of immune suppressing agents and immunomodulators is crucial in suppressing inflammation and possibly preventing fibrosis and irreversible damage. Although the natural progression of IgG4-RD is poorly understood, advanced fibrosis may portend a decreased response to medical therapy.

IgG4-RD is a rare systemic fibroinflammatory condition that can be challenging to diagnose and can mimic solid tumors. If there is a strong suspicion for malignancy with a negative biopsy, clinicians should consider staining for IgG4-RD.
An Uncommon Guest in the Common Bile Duct

Authors: Omeed Alipour MD, Doug Hutcheon MD, and Patrick Sarte MD

Introduction: A 40 year old Mexican man with presumed alcohol cirrhosis and chronic anemia who recently immigrated from Oaxaca, Mexico, where he worked on a farm, was admitted with worsening chills, myalgias, dyspnea, and anemia.

Case Presentation: His exam was notable for right upper quadrant abdominal tenderness. Laboratory studies were remarkable for hemoglobin of 3.6 g/dL, MCV 55 fL, alkaline phosphatase 231 IU/L, and a normal white blood cell count with 23% eosinophils. Due to the anemia, an esophagogastroduodenoscopy and colonoscopy were performed but did not identify a source of bleeding. An MRCP identified an ill-defined mass extending along the common bile duct. Due to concern for cholangiocarcinoma, an ERCP was performed and revealed live mobile flat worms in the common bile duct. Five organisms were removed and identified as Fasciola hepatica, confirmed by CDC pathogen identification.

Triclabendazole, the agent recommended for treatment of human fascioliasis, is not available in the US except under CDC investigational protocol. The infectious disease team contacted the CDC and FDA to obtain the medication. After the FDA and our institution approved the treatment, 1g triclabendazole was administered in two doses after a fatty meal to increase absorption. The patient was discharged the following day without complication.

The patient was seen in clinic 12 weeks after discharge. His hemoglobin improved to 13.2 g/dL and eosinophilia decreased to 7.3%. He felt well and no longer complained of dyspnea or abdominal pain.

Discussion: Human fascioliasis affects over 2.5 million people worldwide. It is found mainly in sheep raising areas in South America, Mexico, Africa, China, and the Middle East, with rare cases reported in the United States. Though sheep are the definitive host for Fasciola hepatica, a snail vector is required, and humans can be infected through contaminated fresh water or ingestion of contaminated uncooked vegetables such as watercress, the suspected source of infection in our patient. The parasite obstructs the common bile duct mechanically or via hyperplasia and hypertrophy of duct epithelium. MRCP can show findings suggestive of cholangiocarcinoma and patients often present with symptoms concerning for biliary obstruction or cholangitis. However, direct visualization remains the definitive method for diagnosis. Triclabendazole is the most commonly reported successful treatment of this disease; it affects the juvenile and adult forms of the fluke, disrupting microtubule function in the organism. Use of bithional has also been reported. There is no FDA approved treatment that can be used in the US without request and investigational protocol.

Human fascioliasis causes severe microcytic anemia. It can lead to inflammatory changes and fibrosis, likely contributing to cirrhosis. In our case, the patient denied chronic alcohol abuse and only reported intermittent alcohol abuse. This suggests that human fascioliasis may lead to cirrhosis even with moderate alcohol intake.

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High Index of Clinical Suspicion remains Paramount to the Diagnosis and Treatment of Necrotizing Fasciitis, Regardless of LRINEC Score

Authors: Ghazal Naz Bahri, MD, Syung Min Jung, MD, FACP

Introduction: Early diagnosis is missed in up to 90% of Necrotizing Fasciitis (NF) cases. The Laboratory Risk Indicator for Necrotizing Fasciitis (LRINEC) score has been introduced as a diagnostic tool for NF. We present two cases of NF with low LRINEC score where in one case, a delayed surgical debridement resulted in mortality.

Case Presentation:

CASE 1: A 60-year-old male patient presented with left arm pain where he injected heroin 3 days prior to admission. He had a low grade fever and other vital signs (VS) were within normal range (WNR). His left upper arm was swollen, warm, hard and tender to touch. No fluctuation or skin changes were noted. His laboratory findings included WBC 13/mm3, Na 132mmol, Lactate 2.3mmol/L, high CRP 6.72mg/dl and CPK 4600unit/L. Given the low LRINEC score of 4, surgical consultation recommended supportive treatment with intravenous antibiotics. His condition worsened with a rise in WBC, CPK and creatinine. On hospital day (HD) 4, he had cardiac arrest. Bedside debridement in the ICU confirmed NF. Patient ultimately passed away from septic shock.

CASE 2: A 45-year-old male patient with pre-diabetes admitted with the infected left leg. His initial vital signs were WNR. His right thigh was erythematous, warm, hard and painful to touch. Skin showed grayish discoloration. His laboratory result showed WBC 22/mm3, bands 36% and elevated CRP. LRINEC score was 5. NF was suspected clinically and debridement was performed on HD 2. Post-operative pathology confirmed NF. The wound culture grew Streptococcus pyogenes. He was discharged home on HD 7.

Discussion: NF is a potentially life threatening emergency condition that causes destruction of soft tissue and fascia. NF leads to a high adverse outcome if not intervened early. LRINEC score has been shown to be a useful adjunct to diagnosing NF early, however only if the score is high (>6).

NF should be highly suspected in any patient with comorbid conditions such as diabetes, liver cirrhosis, IVDA or any immune suppressed condition. Early phase of NF shows severe pain beyond the apparent area of skin, erythema, swelling, induration and fever. The lesion can progress and develop hemorrhagic bullae, crepitus, skin anesthesia, dusky discoloration.

Low LRINEC scores do not exclude NF, and ultimately its diagnosis remains clinical. Both our cases had a low LRINEC score however, high suspicion and early intervention prevented morbidity and mortality in CASE 2.

Conclusion: Recognition of NF and early debridement is the most important determinant of outcome in NF regardless of LRINEC score. A multidisciplinary discussion on the above cases has led to creating a protocol and training our ED and surgical providers on clinical diagnostic clues and need for urgent surgical intervention for any suspected case of NF.

References


Authors: Bakshi, A

Introduction: Klebsiella pneumoniae infections are often seen in patients with co-morbidities or immunosuppression. In recent years, Klebsiella pneumoniae has been the most common pathogen causing pyogenic hepatic abscess with highest incidence in East Asia, particular

Case Presentation: A 41-year-old Caucasian male with no co-morbidities presented with right upper quadrant and left flank pain, fever, chills, nausea, anorexia for 1 week and pain with decreased vision of the left eye for 5 days. On examination, he was afebrile with abdominal tenderness in the right upper quadrant and left costovertebral angle tenderness. Left eye exam revealed conjunctival hyperemia, minimal light perception, corneal haziness and hypopyon. Pertinent laboratory results revealed creatinine 1.08mg/dl, total bilirubin 5.5mg/dl, AST 76U/L, ALT 87U/L, ALP 368U/L and white cell count 18.4x10^3/ul. Urinalysis demonstrated pyuria and bacteriuria. CT scan of abdomen and pelvis demonstrated left kidney pyelonephritis and abscess and a septated abscess in the liver measuring approximately 4.8x4.5x6cm. CT of the facial bones and MRI of the head showed circumferential preseptal and postseptal edema compatible with endophthalmitis with a phlegmon. Blood and urine cultures were positive for Klebsiella Pneumoniae. Urgent ophthalmology consult was obtained and patient was deemed as an unfavorable candidate for vitrectomy or intravitreal antibiotics. Patient was treated with intravenous ceftriaxone 2g daily for 6 weeks which resulted in decrease in size of liver abscess. Patient was recommended to continue ciprofloxacin 500mg orally twice daily until complete resolution of abscess and future enucleation.

Discussion: K. pneumoniae liver abscesses often appear more solid with less purulent aspirate than other etiologies. Staphlococcus aureus, Streptococcus pyogenes and Candida species are alternative causes of pyogenic liver abscesses. A definitive diagnosis can be made with positive blood or aspirate cultures as there is usually concurrent bacteremia. Although rare, metastatic infection such as endophthalmitis, pulmonary abscesses, and CNS involvement are seen in pyogenic liver abscess. Second or third generation cephalosporins and fluoroquinolones are the mainstay of treatment. Carbapenems are used for extended spectrum beta-lactamase (ESBL) strains. Treatment duration is between four and six weeks depending on abscess size and metastatic progression. Regular follow-up imaging is recommended to monitor complete resolution. Endophthalmitis warrants early detection and initiation of antibiotics and prompt ophthalmology consultation as it may lead to vision loss if not treated urgently. Vitrectomy and intravitreal antibiotics may be required to prevent progression and enucleation or evisceration may be considered if medical treatment fails.

The patient presented in the case is Caucasian without any identifiable risk factors suggesting that incidence of Klebsiella liver abscess is increasing worldwide and should be at the forefront of differential diagnosis irrespective of geographical preponderance, race or other comorbidities.

References


The Story of a Good Cough

Authors: Patrick Chan, MD, PGY-2, Doug Hutcheon, MD and Patrick E. Sarte, MD, MS, FAAP, FACP

Introduction: Chronic cough is a common outpatient chief complaint that has a broad differential. Most of the time, routine work up is sufficient to diagnose the etiology; however, for refractory cases, it is important to broaden differentials as there can be clinically significant treatment options.

Case Presentation: Patient is a 57-year-old male with history of resected thymoma two years prior who presented to pulmonary clinic for chronic cough of one year. Patient had daily productive cough of yellow-green sputum, which would resolve with antibiotics. Review of systems was negative for tuberculosis, allergy symptoms, or GERD symptoms. Patient was not on an angiotensin-converting-enzyme (ACE) inhibitor. His social history is significant for emigration from Philippines 10 years ago, prior work exposure to pesticides and cleaning solutions, and 30-pack year smoking history. However, patient had no recent travel and no exposure to pets or birds. Patient was tried extensively on OTC and prescription therapies for cough, allergy, and GERD with no improvement in symptoms. Patient even had referral to ENT specialist to evaluate for etiology of cough, who reported an unremarkable exam. Patient was eventually referred to pulmonary clinic, and was presumptively diagnosed with worsening COPD. On subsequent visits, despite medical optimization, patient still had symptoms, and was eventually worked up for immunodeficiency given history of thymoma. Patient’s work up was significant for low immunoglobulins - IgM, IgG, IgA. Patient was then diagnosed with Good’s syndrome, and was started on intravenous immunoglobulin (IVIg) with good initial response, with a one year period with no hospitalizations or emergency department visits. However, patient has since relapsed and has had several hospitalizations, with one being for severe multifocal pneumonia.

Discussion: Good syndrome is a rare entity, which is characterized by an association between thymoma and acquired immunodeficiency. Due to its immunodeficiency, the clinical presentation is often with recurrent sinopulmonary infections. Furthermore, the immunodeficiency does not resolve after thymectomy, and the mainstay of treatment for patient with Good’s syndrome is IVIg replacement. However, despite IVIg replacement, patients may still have progression of disease and have refractory infections. Our patient followed the classical clinical presentation, with recurrent infections after thymectomy that initially responded to immunoglobulins. This case illustrates the importance of taking a detailed history and placing it in clinical context. The patient has been followed as an outpatient in primary care clinic and was treated several times for recurrent pneumonia. It was natural to presume that our patient had COPD exacerbation given his smoking history; however, the history of thymoma and recurrent pneumonias pointed toward a diagnosis of Good’s syndrome, and the patient was able to receive IVIg that allowed the patient to be symptom free for a one year period.

References

Recurrent Priapism – A Peculiar Presentation of Chronic Myeloid Leukemia

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Introduction: Priapism is persistent abnormal erection of the penis without accompanied sexual desire or stimulation. Although priapism may often be idiopathic, the condition can be secondary to various causes such as sickle cell disease, leukemia, trauma, thromboembolic disease, medications, and drugs. The following case illustrates recurrent priapism as a rare presenting symptom of chronic myeloid leukemia (CML).

Case Presentation: A 20-year-old male was referred to urology for an eight-week history of new onset weekly painful priapism, with episodes typically self-resolving and with the longest episode lasting 6 hours. During these eight weeks, the patient had visited various emergency departments for evaluation and each time presented after resolution of his priapism. The only workup completed during this time included a negative sickle cell screen, with pseudoephedrine prescribed as needed. He was eventually referred to urology, where a complete blood count (CBC) revealed a white blood count (WBC) of 288×10³/mcL concerning for hematological malignancy. There was no trauma, intake of new medications, drug or tobacco use, exposure to radiation, bleeding, fever, or chills. Review of systems was positive for night sweats, lightheadedness, fatigue, nausea, headache, and decreased appetite. Patient denied any family history of hematological disorders. Careful history taking revealed patient worked in the vehicle industry and had significant exposure to powder coating and various chemicals including benzene.

Physical exam revealed no conjunctival pallor, lymphadenopathy, jaundice, or ecchymoses. Abdominal examination demonstrated no tenderness nor splenomegaly. His penis was not erect. Repeat CBC showed WBC count: 314×10³/mcL, hemoglobin: 9.5 g/dL, and platelets: 402×10³/mcL. Differential cell count revealed 24% neutrophils, 9% bands, 13% metamyelocytes, 18% myelocytes, 19% promyelocytes, 7% eosinophils, and 3% basophils. Uric acid was 6.9 mg/dL and lactic dehydrogenase was 792 IU/L. Renal, hepatic, and coagulation profiles were all normal.

A peripheral blood smear confirmed myeloid leukocytosis with left shift without blasts. Patient was started on allopurinol and hydroxyurea. BCR/ABL p210 transcript was confirmed on peripheral blood, and bone marrow biopsy was consistent with CML in chronic phase. Cytogenetics confirmed the presence of the Philadelphia chromosome. He was commenced on imatinib 400 mg daily. One month after initiation of therapy, his WBC count was 6.1×10³/mcL, and both hydroxyurea and allopurinol were discontinued. He has not had any recurrent priapism.

Discussion: In this case, the patient would have received a more prompt diagnosis of CML had a CBC been checked during his multiple emergency department visits in the initial eight-week timeframe. The history of benzene exposure is interesting as some epidemiological studies suggest an association between occupational benzene exposure and CML. Priapism is a rare presenting feature of CML, and the mechanism is related to hyperleukocytosis with aggregation of leukemic cells in the corpora cavernosa and the dorsal veins of the penis. Although an unusual presentation, early recognition with proper workup is critical to diagnosis and initiation of appropriate therapy.
Treating Cutaneous Metastasis of Breast Cancer with Topical Imiquimod

Authors: Nguyen, Anthony MD, Chong, Esther G. MD, Shaheen, Shagufta MD and Mirshahidi, Hamid MD

Introduction: Cutaneous metastasis of breast cancer carries a poor prognosis, invokes a poor quality of life, and increases mortality by increasing one’s risk of bleeding and infection. Current therapies include systemic chemotherapy, surgical resection and radiation, all of which are invasive and can have toxic side effects.

Case Presentation: A 50-year-old African-American woman was diagnosed with left breast ductal carcinoma in situ (DCIS) and was treated with left total mastectomy. 6 years later, she was diagnosed with stage IV breast adenocarcinoma (ER/PR positive and HER-2 negative) with recurrence and metastasis in multiple lymph nodes. She then completed chemotherapy with concurrent radiation therapy with good clinical and radiographic response. 4 years after breast cancer recurrence, she noted skin hardening, pain and hyperpigmentation over her left chest wall, with increasing lymphatic masses in her left neck and left supraclavicular area. PET-CT demonstrated cervical, supraclavicular, mediastinal and axillary lymphadenopathy with increased uptake in the left upper lung, sternum, and left chest wall consistent with breast cancer recurrence and metastasis. Despite chemotherapy, skin lesions continued to worsen with burning pain and became hard with frequent ulcerations.

To treat her metastatic skin lesions to the upper left chest wall and left supraclavicular area, she was started on topical imiquimod 5% cream to her skin lesions twice per day for 5 days a week. Within 4 months of starting topical imiquimod cream, the lesions decreased in size, thickness, and pigmentation, with resolution of ulceration as evidenced by serial photographic and clinical documentation. After improvement of skin lesions within 4 months of use, she started to use imiquimod on an as-needed basis, according to her symptoms and size of her lesions while undergoing systemic chemotherapy treatment.

Discussion: Our case demonstrates the longest interval between initial diagnosis of the patient’s primary breast cancer and eventual cutaneous metastasis (10 years) with successful treatment with topical imiquimod. This case adds to the growing body of literature demonstrating that topical imiquimod cream is a treatment option for cutaneous metastasis of breast cancer. Imiquimod has been described in recent literature to have anti-tumor properties and the ability to induce a pro-immunogenic tumor microenvironment. The regression and improvement of our patient’s skin lesions after starting on imiquimod was clinically significant and further demonstrates the efficacy of topical imiquimod. It holds excellent promise as a treatment option for cutaneous metastasis of breast cancer in conjunction with systemic immunomodulatory therapies and chemotherapy for symptomatic and cosmetic management.
Say N2O to Drugs: A Unique Presentation of Nitrous-Oxide Induced B12 Deficiency

Authors: Maralee Kanin, MD., Lisa Tsang, MD., and Julien Nguyen, MD

Introduction: Vitamin B12 deficiency is associated with a wide array of neurologic, psychiatric, hematologic, dermatologic, cardiovascular and gastrointestinal manifestations. Neurologic manifestations include: paresthesia, ataxic gait and subacute combined degeneration of the spinal cord. Vitamin B12 deficiency is typically the result of GI tract abnormality such as malnutrition, malabsorption, or gastrectomy. We present an atypical cause of symptomatic B12 deficiency due to substance abuse.

Case Presentation: A 19 yo woman with no past medical history presented with one month of progressive bilateral hand and feet numbness and ataxic gait. Her history was unremarkable, she denied abdominal operations, consumed meat, and initially denied substance abuse. Further discussion revealed “huffing” of 20 gallons of nitrous oxide (N2O) on weekends for about 6 months. Her physical exam was notable for diminished sensation to light touch, pinprick and vibratory sense on dermatomes C5-T1 and L5-S3. She was also noted to have a broad-based gait requiring a walker, and her Romberg test was positive. Initial studies revealed a mild macrocytic anemia (hemoglobin 11.0/ MCV 96), HIV negative, RPR negative, and SPEP/UPEP negative. Further labs include B12< 146, folate 13, MMA 16,420, Homocysteine 86.6. MRI of the brain revealed non-enhancing long plaques extending from the C1 level through at least the superior margin of T4. Drug cessation was counseled. She was started on B12 injections. B12 levels returned to normal and symptoms completely resolved by post hospitalization follow-up at 2 months

Discussion: B12 is an essential cofactor for the conversion of homocysteine to methionine and MTHFR (methylene-tetrahydrofolate reductase) to THF (tetrahydrofolate), which are responsible for the production of DNA and myelin, respectively. The B12 enzyme facilitating these reactions has a central cobalt in the 1+ state, and nitrous oxide oxidizes the cobalt to the 3+ state rendering the B12 enzyme inactive. Thus, nitrous oxide effectively slows DNA and myelin synthesis, which can lead to subacute combined degeneration (SCD). SCD is a degeneration of the dorsal and lateral white matter columns of the spinal cord. The result is a slowly progressive weakness, sensory ataxia, and paresthesias. If left untreated can lead to spasticity, paraplegia, and incontinence. N2O can be easily obtained in whipped cream canisters, air duster for computers and from octane booster canisters at auto repair shops. Our patient worked in an auto repair shop, which is how she obtained her drug of choice. Subacute combined degeneration is a complication of severe B12 deficiency. Recreational usage of Nitrous Oxide is a rare cause of B12 deficiency. In young, otherwise healthy patients, thorough substance history should be obtained. N2O cessation and B12 supplementation is the treatment.

References

Crystalizing Colitis

Authors: T. Lai, D.O.; B. Barrows, D.O.; M. Salehpour, M.D.; A. Frugoli D.O., Community Memorial Health Sciences

Introduction: End-stage renal disease is associated with multiple metabolic and electrolyte derangements. Hormonal and electrolyte imbalance of calcium, phosphorus, and parathyroid hormone (PTH) can result in short term complications such as calciphylaxis and long term sequelae such as renal osteodystrophy. As a result, many patients have strict dietary constraints and despite compliance, phosphate binders such as calcium acetate and/or sevelamer carbonate (Renvela) are also needed to treat secondary hyperparathyroidism. This case vignette describes an under recognized adverse effect of a phosphate binder, Renvela induced colitis.

Case Presentation: A 47 year old male with insulin dependent diabetes complicated by end-stage renal disease presented with a two day history of abdominal pain with associated nausea. Of note, he presented one month prior with similar symptoms. During his previous admission he underwent evaluation with CT imaging, followed by colonoscopy and was found to have numerous circumferential lesions not consistent with inflammatory bowel disease, but suspected to be related to ischemia. He was provided supportive care and non-operative management and was able to be discharged home. He returned with similar symptoms a month later. He was empirically started on antimicrobials and underwent additional evaluation with repeat imaging using CT mesenteric angiogram that demonstrated colitis in distal ascending, transverse and proximal descending colon with pericolic stranding, but no evidence of atherosclerotic disease. He again underwent repeat colonoscopy that re-identified 3 areas of circumferential ulceration in ascending/hepatic flexure, splenic flexure and descending colon. Pathology report noted necrotic tissue, ulcerative debris with non-polarizing crystalline material from biopsies taken from aforementioned areas of ulceration. Evaluation for additional causes including cardioembolic events and vasculitis were bland. Review of prior pathology and current biopsy specimens demonstrated crystalline material that is most likely related to Renvela use. The patient was discontinued off of Renvela and transitioned to calcium carbonate. His symptoms improved with cessation of Renvela and initiation of its substitute.

Discussion: This vignette describes a rare adverse effect of sevelamer carbonate therapy. Review of the literature show a limited number of reported cases. Additionally, cases of renvela resin related necrosis can be misidentified with cases of Kayexalate or bile acid sequestrants, as all represent medication resin resulting in colonic necrosis. Pathologist are able to distinguish Kayexalate’s morphology from Renvela as, it is rectangular with internal demarcation described as “fish scales”, with a very characteristic hue that was described by Gonzales et al & Swanson et al. Given the very small incidence, no definitive pathophysiology has been confirmed and only limited reports of pathologic identification exists. We offer review of the pathologic tissue finding, as well as a proposed mechanism of tissue injury.

References

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Pemetrexed-Induced Stress Cardiomyopathy

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Introduction: Since its description in 1990, stress cardiomyopathy (SC), or takotsubo cardiomyopathy, has been associated with physical or emotional stress. However, medication induced SC may be an important consideration in cancer patients receiving chemotherapy. We present a case of SC associated with pemetrexed use. To our knowledge, this is the first reported case of pemetrexed-induced SC.

Case Presentation: A 79-year-old woman with stage IV lung adenocarcinoma presented with substernal chest pain. Three days prior, she was started on pemetrexed (Alimta). Other medications included ondansetron, simvastatin, metformin, and lisinopril. She denied use of sympathomimetic drugs, recent infection, emotional, or physical stress.

Troponin was elevated at 1.35 nanograms/milliliter and electrocardiogram (ECG) showed ST elevations in the lateral leads. Cardiac catheterization revealed apical hypokinesis with an ejection fraction (EF) of < 20% and no obstructive coronary artery disease.

Pemetrexed was identified as the inciting factor and discontinued. Repeat echocardiogram two days after cardiac catheterization revealed an improved EF of 45-50%. She was medically optimized with captopril, furosemide, and bisoprolol with resolution of symptoms.

Discussion: SC is characterized by apical hypokinesis usually incited by physical or emotional stress with resultant transient systolic heart failure. Patients present with acute ischemic ECG changes and elevated troponin suggestive of acute coronary syndrome (ACS), but without obstructive coronary artery disease.

Pemetrexed is a folate analog metabolic inhibitor. The drug inhibits enzymes essential to the folate-dependent metabolic pathway in the process of DNA replication. It is theorized that pemetrexed may cause cardiotoxicity via production of reactive oxygen species due to elevated homocysteine levels similar to methotrexate, another folate antagonist.

Since 2000, 27 cases of cancer-treatment related SC have been reported. 19 cases were due to chemotherapeutic agents, 6 from monoclonal antibody therapy, and 2 from tyrosine kinase inhibitor treatment. Time course of onset varied from immediate to 6 weeks after treatment with most cases occurring within 6 days. Of note, 3 of the 27 cases were rechallenged with the suspected agent and 2 of the 3 cases developed myocardial infarction with rechallenge. The pathogenesis of cancer-therapy related SC is thought to occur via the drugs’ direct cardiotoxic effects from the generation of free radicals rather than a stimulated release of catecholamines.

This case raises concern for the association of pemetrexed and SC. Early recognition and discontinuation is critical to its resolution. The offending agent should not be rechallenged in a patient who develops SC. Instead, alternative therapies should be considered.
An Unsolved Mystery: Idiopathic Hypereosinophilic Syndrome Masquerading as Recurrent Ascites

Authors: Ni Mo, DO; Stan Amundson, MD

Introduction: Hypereosinophilic syndrome (HES) is a group of clinical syndromes characterized by hypereosinophilia (absolute eosinophil count >1500/mcL) and evidence of end-organ injury attributable to eosinophilia. While some forms of HES have identifiable etiologies, a significant number of cases have undefined causes.

Case Presentation: A 48-year-old female with past medical history of hyperthyroidism and anxiety presented to the emergency room with several weeks of abdominal pain, bloating, and early satiety. She had similar symptoms in 2006 with no identifiable cause. Her only medications are sertraline and propylthiouracil. She has no known allergies. Physical exam was notable for a moderately distended abdomen with shifting dullness to percussion. Laboratory data was significant for WBC count of 15,600/mcL with 54% eosinophils (absolute eosinophil count 8460/mcL). CT abdomen showed diffuse thickening of distal stomach with mild abdominal ascites. An extensive infectious workup was negative for parasitic etiologies. Esophagogastroduodenoscopy revealed grossly normal findings with normal gastric biopsy results. She was discharged on an empiric course of ivermectin and albendazole.

Patient was re-admitted one week later with worsening abdominal distension and dyspnea. Laboratory data showed persistent eosinophilia (absolute eosinophil count 7270/mcL) and troponin of 0.488 ng/mL. A repeat CT abdomen showed diffuse small bowel thickening with large amount of ascites. Paracentesis was performed, and fluid analysis revealed WBC count 5320/mcL with 93% eosinophils and SAAG of 0.5, without evidence of peritonitis or malignancy. Echocardiogram and cardiac MRI were both normal. Bone marrow biopsy revealed hypercellular marrow with 45% eosinophils, but normal female karyotype with no abnormalities detected in PDGFRA, PDGFRB, FGFR1, and BCR-ABL genes by FISH analysis. Additional studies only pertinent for mildly elevated IgE and serum IL-5 levels. She was treated with high dose methylprednisolone, which resulted in rapid resolution of eosinophilia. She was discharged home on a prednisone taper.

Discussion: The prevalence of HES is estimated to be 0.6 to 6.3 per 100,000. While it is a rare group of disorders, it can potentially result in life-threatening complications from eosinophilic infiltration and damage to organs, particularly with cardiac and neurologic involvement. Our patient presented with troponinemia during the second hospitalization. Though the echocardiogram was normal, the cardiac MRI showed subepicardial signal enhancement at the inferior septum with undetermined significance, which may be compatible with subclinical myocarditis. The patient also had evidence of gastrointestinal tract thickening on two separate CT scans. While biopsy results from EGD did not show eosinophilic involvement, it is possible the samples obtained were not deep enough. Treatment of HES depends on the subtype, but it generally responds well to glucocorticoids. Though no specific etiology was identified for our patient’s eosinophilia, she improved remarkably with high dose steroids. Other treatment modalities include agents targeting IL-5, and are undergoing clinical trials to investigate their efficacy in treating HES.

References

Recognizing Leser-Trelat Sign as Primary Presentation of Gastrointestinal Malignancy

Authors: Hannah Moser, MD; Andy Liu, MD; Ingeborg Schafhalter-Zoppoth, MD

Introduction: Leser-Trelat sign, a paraneoplastic dermatosis, is characterized by the rapid onset of seborrheic keratoses associated with an occult malignancy. Given the increased incidence of seborrheic keratoses in elderly patients, some skepticism exists about the sign's validity. However, knowledge of this sign can lead to fast, non-invasive diagnosis and better outcomes for patients. We report a series of two cases which demonstrate how a primary care physician’s prior knowledge of the sign can allow for more rapid diagnosis and patient-centered outcomes.

Case Presentation

Case #1:

A 73-year-old man with a history of treated hepatitis C infection presented initially with the sudden appearance of multiple hyperpigmented lesions, and was later found to have gastric adenocarcinoma.

The patient first observed a single, flesh-colored, non-tender lesion on his lower back. Six weeks later, he had developed multiple hyperpigmented lesions across the same area, as well as axillary acanthosis nigricans. He denied weight loss, abdominal pain, or dysphagia. On routine screening ultrasound for hepatocellular carcinoma, a pancreatic mass was identified. Follow-up CT scan revealed thickening of the gastric fundus as well as omental adenopathy. On endoscopy, a large, fungating, non-circumferential mass was found on the lesser curvature of the stomach; biopsies were consistent with HER-2/neu negative invasive gastric adenocarcinoma.

The patient chose to undergo aggressive treatment at first, and eventually transitioned to hospice care.

Case #2:

Several months after his death, a 93-year-old woman presented to the same physician with the sudden appearance of numerous hyperpigmented skin lesions.

The patient noted the abrupt development of pruritic lesions across her back, neck and chest. She denied weight loss, abdominal pain, nausea, or changes in bowel function. Biopsies of the skin lesions revealed seborrheic keratosis. However, given the physician’s prior experience with Leser-Trelat, the potential for an underlying malignancy was addressed. CT of the abdomen revealed asymmetric wall thickening involving the proximal to mid-ascending colon, raising concern for colon cancer. Six hypodense liver lesions were suspicious for metastatic disease. The patient decided to pursue single-agent palliative chemotherapy with capecitabine for several weeks, and then transitioned to hospice care. She passed away shortly thereafter at home with her family.

Discussion: In conclusion, this case series illustrates the value of physician knowledge of Leser-Trelat sign in a primary care office. Malignancy was diagnosed fast and non-invasively before symptoms occurred. The primary care physician was able to start the conversation of end-of-life decisions early. Consequently, the patient was able to attain hospice care and receive extra support at the end of her life without expensive and upsetting ED visits and hospitalizations.
Pediculosis: An uncommon cause of severe iron-deficiency anemia

Authors: Lowell Thorndike Nicholson, MD, Ying Li, MD, PhD, Dolores Shoback, MD

Introduction: Iron-deficiency anemia is the most common cause of anemia worldwide and is routinely encountered in clinical practice. In resource-rich countries, iron-deficiency is most often caused by chronic blood loss, malabsorption, or reduced dietary intake. Although occult bleeding is frequently found in the gastrointestinal tract, here we present a case of severe iron deficiency anemia due to heavy louse infestation.

Case Presentation: A marginally-housed 59-year-old male with a history of well-controlled schizophrenia and chronic anemia was referred to the hospital by his primary care physician after he was found to have a hemoglobin of 5.7 g/dL. During the 12 months before his hospital admission, the patient had been seen regularly in the outpatient setting and his hemoglobin had ranged between 9 and 12 g/dL. Four months prior to his current presentation, fecal occult blood testing was performed and was negative. The patient’s anemia was presumed to be due to reduced dietary intake and he was started on oral iron supplementation. On admission to the hospital, the patient reported feeling sluggish but denied any overt bleeding or melena. Physical exam revealed numerous lice on the patient’s body and clothing as well as multiple excoriated ulcerations on the torso, back, and extremities. Initial laboratory testing was notable for peripheral eosinophilia, an elevated erythrocyte sedimentation rate, and severe iron deficiency with a very low absolute reticulocyte count. Fecal occult blood testing was negative. Urinalysis, thyroid stimulating hormone, and results of kidney and liver function testing was normal. Serum protein electrophoresis and H. pylori testing were negative. Lice were collected and samples were sent to pathology which identified Pediculus spp. The patient was transfused with two units of red blood cells and received intravenous iron supplementation for 5 days. His louse infestation was treated with several rounds of topical permethrin. At the time of discharge, the patient’s hemoglobin had improved to 8.5 g/dL with an appropriate reticulocytosis. The patient’s social worker was contacted and agreed to help the patient thoroughly clean his home and belongings. He was recently seen in the outpatient setting for follow-up at which time his hemoglobin had improved to 10.9 g/dL and his peripheral eosinophilia had resolved.

Discussion: The body louse (Pediculus humanus humanus) feeds on human blood and chronic infestation can result in clinically significant blood loss and iron-deficiency anemia. Louse infestation is common in marginally-housed patients and those with severe mental illness. As this case illustrates, pediculosis-associated iron-deficiency may be an overlooked cause of anemia in these patient populations.
Leave No Stone Unturned: Recurrent Pyogenic Cholangiohepatitis Masquerading as Choledocholithiasis, Cholangitis, and Cirrhosis

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Introduction: Recurrent pyogenic cholangiohepatitis is endemic in Southeast Asian populations and is an underrecognized cause of hepatobiliary obstruction in western countries. It is exceedingly rare in populations from other ethnic groups. Here, we present a case of a patient with recurrent pyogenic cholangiohepatitis to illustrate the diagnostic challenges of this disease.

Case Presentation: A 74-year-old woman from Mexico with chronic liver disease and prior hepatitis B infection presented with several months of worsening jaundice, lower extremity edema, and fatigue. One month prior, the patient was prescribed herbal supplements. On admission, initial labs were notable for transaminitis with a cholestatic pattern: AST 156, ALT 44, ALP 727, Tbili 8.1. Lab tests for acute hepatitis were unrevealing. Abdominal ultrasound demonstrated choledolithiasis without evidence of cholecystitis and common bile duct dilation (CBD) 7 mm. Abdominal CT demonstrated capsule nodularity of liver concerning for cirrhosis and diffuse dilation of intrahepatic bile ducts with hepatolithiasis and choledocholithiasis. Our initial differential diagnosis included choledocholithiasis, drug-induced cholestasis, malignancy, primary biliary cirrhosis, and primary sclerosing cholangitis. Endoscopic Retrograde Cholangiopancreatography (ERCP) showed markedly dilated left intrahepatic ducts with innumerable filling defects as well as dilated common hepatic duct and common bile duct which underwent balloon sweeps with copious amounts of sludge and stone material removed. The patient was diagnosed with recurrent pyogenic cholangiohepatitis based on the degree of stone burden and findings of intrahepatic and extrahepatic bile duct dilation seen on ERCP.

Recurrent pyogenic cholangiohepatitis is a disease characterized by recurrent liver inflammation due to intrahepatic and extrahepatic obstruction from de novo biliary stone formation composed of calcium bilirubinate in contrast with cholesterol stones with cholelithiasis and choledocholithiasis. The disease is classically seen in patients from Southeast Asia through unknown mechanisms. Previous epidemiological studies have suggested that parasitic infections such as Clonorchis sinensis and Ascaris lumbricoides may play a role in the pathogenesis of this disease. Stool ova and parasites were negative in our patient. Management of this disease involves antibiotics, biliary decompression, and possible surgery for severe, recurrent disease. Our patient was treated with seven days of amoxicillin/ clavulanate and scheduled for definitive biliary decompression with ERCP with SpyGlass for electrohydraulic lithotripsy. Future surgical options for this patient would include hepatic ductal resection and biliary-enteric anastomosis.

Discussion: This case was notable for an uncommon presentation of an uncommon disease and illustrates that recurrent pyogenic cholangitis may occur in patients not from classic endemic regions or ethnicities. It is possible that there is an under diagnosis of this disease in the US. Our case also highlights that recurrent pyogenic cholangitis may be a cause of secondary biliary cirrhosis. In conclusion, clinicians should broaden their differential diagnosis to include recurrent pyogenic cholangitis in patients with cholestatic liver disease with multiple hepatobiliary stones.
CALIFORNIA CLINICAL VIGNETTE POSTER FINALIST - CHARNJEET SANDHU, MD

An Unusual Case of Schmidt’s Syndrome with Severe Hemodynamic Crisis Requiring ECMO

Authors: Charnjeet S. Sandhu MD, Natasha Singh MD, Department of Internal Medicine, UCSF-Fresno

Introduction: In Western countries, the primary cause of adrenal insufficiency is autoimmune disease, which accounts for 70 - 90% of cases. About 50 – 65% of patients with autoimmune adrenal disease have other autoimmune endocrine disorders referred to as Autoimmune Polyglandular Syndrome (APS). These disorders are divided into APS type 1 and type 2. Herein, we present an interesting case of newly diagnosed Schmidt’s syndrome (APS II) complicated by significant hemodynamic instability that was refractory to pressors and requiring Venous-Arterial Extracorporeal Membrane Oxygenation (VA-ECMO) to achieve hemodynamic stability.

Case Presentation: A 31-year-old Hispanic male with recent diagnosis of hypothyroidism started on Levothyroxine one month ago was transferred from his primary’s office to emergency room (ER) for hypoglycemia and severe hypotension. One-week prior, patient had been experiencing severe fatigue, weight loss, nausea, and non-bloody emesis with each meal along with skin darkening. Vital in ER: blood pressure 64/38, pulse 82/min and oral temperature 35C. He was mentating normally, thin, there was evidence of hyperpigmentation in the oral mucosa (including the gums and sublingual area), face, and nails. Laboratory evaluation revealed a glucose 65mg/dl, sodium 121mmol/l, TSH 6.4IU/ml, free T4 1.54ng/dl, and random serum cortisol was <0.40ug/dl. Arterial blood gas showed severe metabolic acidosis with pH of 7.14, PCO2 21, and pO2 of 101mmHg on 100% oxygen. Bedside echocardiogram showed a small pericardial effusion without tamponade physiology and ejection fraction of 50-55%. He received aggressive fluid resuscitation and was initiated on stress doses of hydrocortisone. Despite adequate resuscitation and maximum vasopressor support, he remained hypotensive. Shortly patient suddenly developed cardiac arrest with PEA. Return of spontaneous circulation was achieved with ACLS protocol. Patient was transferred to operating room and was placed on VA-ECMO support. On day three, patient was successfully weaned off VA-ECMO. Further workup confirmed a diagnosis of APS type 2, with positive anti-peroxidase antibodies (267IU/ml) supporting the diagnosis of Hashimoto’s thyroiditis.

Discussion: The diagnosis of APS-2 requires two or more of the following: primary adrenal insufficiency, thyroid disorder (Graves or autoimmune thyroiditis), type 1 diabetes mellitus and/or primary hypogonadism. The most common clinical combination is the association of Addison disease and Hashimoto thyroiditis. Review of outpatient records revealed that patient was started only on levothyroxine. Initiation of thyroid replacement without steroids in this patient with Hashimoto’s thyroiditis may have precipitated an acute adrenal crisis in our patient due to coexisting adrenal insufficiency. This case illustrates the importance of recognizing subtle physical exam findings such as hyperpigmentation as well as initiation of complete workup of associated autoimmune disorders prior to initiating treatment for a commonly seen condition such as hypothyroidism.
Why isn’t he bleeding? Factor VII Deficiency

Authors: Kalbir Singh, MD; Amandeep Gill, MD

Introduction: Factors VII is a vitamin K dependent protein which is produced by the liver and is essential for hemostasis. Factor VII deficiency can be acquired or inherited. Acquired factor VII deficiency can be due to vitamin K deficiency, liver disease, malignancies like Wilm's tumor, sepsis, antiphospholipid antibody, or aplastic anemia; other causes are very rare.

Case Presentation: A 40-year-old Hispanic male with history of drug abuse and long term Vitamin K therapy for presumptive rat poison (d-Con/superwarfarin) poisoning in December 2017, presented from clinic for confusion and supratherapeutic INR. CT head was negative for acute pathology. His lab results were significant for elevated PT at 101 seconds, INR > 10.4, and PTT 26. Patient denied any history of bleeding from gums, nose, GI, or genitourinary. He also denied excess bleeding at time of his hernia repair. Despite months of vitamin K supplementation, patient’s INR and PT remained high since presumed rodenticide poisoning. This prompted evaluation for coagulopathies. Mixing studies were done that were negative for any inhibitor. This was followed by factor deficiency assays, that were significant for factor VII clotting activity <1%. Thus, patient was diagnosed with isolated factor VII deficiency and discharged with INR 3.9 without vitamin K supplementation.

Discussion: PT prolongation suggests a deficiency or inhibition of Factor II, V, VII, X. In approach to abnormally prolonged PT, it is important to obtain history of bleeding during previous surgeries, blood transfusions, or heavy menses. Also history of medication use, herbal products, or drug ingestion would be pertinent. Abnormal coagulation times in patients without bleeding prompts further evaluation with mixing studies. Mixing studies determine the presence of an inhibitor by demonstrating persistent abnormal clotting times when patient plasma and normal plasma is mixed. If no inhibitor is identified on mixing studies, subsequent evaluation done with factor assays to discover factor deficiency.

Factor VII deficiency presents with a wide spectrum of clinical severity that correlates poorly with plasma factor VII levels. Some patients with undetectable levels are asymptomatic, like our patient. In conclusion, it is important to identify underlying factor deficiencies to ensure hemorrhage control when necessary. Apart from antifibrinolytics, FFP, and prothrombin complex, recombinant-activated-FVIIa concentrates have been used in factor VII deficiency induced bleeding. In those without access to factor VII concentrate or recombinant-activated-FVIIa, bleeding can be managed with 4 factor PCC or FFP with care to avoid volume overload. Especially those undergoing minor surgery or minor bleeding are better treated with antifibrinolytic since half-life of factor VII is very short and amount of FFPs required usually causes volume overload.
N2O laughing matter: a case of subacute combined degeneration induced by recreational nitrous oxide inhalation

Authors: Steven Wu, MD; Talar Kavafyan, MD; John Carmody, MD, Huntington Hospital, Department of Internal Medicine

Introduction: Nitrous oxide has long been used for anesthetic purposes, but it is also recognized as a recreational inhalant for its euphoric effects. This case illustrates a patient who presented with inability to walk and was diagnosed with subacute combined degeneration (SCD), as a result of using nitrous oxide. An interesting constellation of signs, symptoms, laboratory abnormalities, and radiographic findings are associated with SCD, and it is important to exclude other differential possibilities.

Case Presentation: The patient is a 32 year-old lady with no medical problems, who presented with difficulty ambulating for the past 1 month. She began to notice intermittent numbness and tingling in her toes and fingertips 7 months ago, which have spread to her calves and hips within the past month. She reported difficulty discerning where her legs were relative to the floor, and her legs became weak to the point that she required a wheelchair. She was 12 weeks pregnant and planned to terminate the pregnancy because of high risk. Her obstetrician referred her to a neurologist, who referred her to the emergency department. She denied any bowel or bladder incontinence, or loss of sensation in the pelvic area. She was not vegan or vegetarian.

Upon further questioning, it was revealed that for the past several months, she had been increasing her use of so-called “whippets,” referring to the nitrous oxide canisters used in refillable whipped cream containers. On examination, she had significant symmetric lower extremity weakness, accompanied by decreased proprioception, vibration sense, and deep tendon reflexes. She was found to have macrocytosis without anemia, hypersegmented polymorphonuclear leukocytes, and vitamin B12 deficiency with elevated homocysteine and methylmalonic acid levels; folate level was normal and intrinsic factor antibody was negative. MRI of the cervical and thoracic spine showed diffuse hyperintensity of the dorsal and central spinal cord.

Neurology was consulted; she was diagnosed with SCD from vitamin B12 deficiency, secondary to nitrous oxide use. She received vitamin B12 supplementation. She required maximum assistance on initial physical therapy evaluation, but she only required minimal assistance to ambulate with a walker when she was discharged home on hospital day 15.

Discussion: This case demonstrates the devastating and potentially irreversible neurologic sequelae due to nitrous oxide use. In particular, it is important to note that nitrous oxide can cause demyelination of the dorsal and lateral spinal cord by irreversibly inactivating vitamin B12, an important metabolite in methylation reactions and DNA synthesis. In fact, it has been documented in similar cases that prophylactic vitamin B12 supplementation in the setting of nitrous oxide use does not always prevent the development of SCD; treatment of SCD with vitamin B12 supplementation unfortunately does not always improve functional recovery, either.

References

Upset stomach: an unusual explanation in a patient with CLL and AIHA

Authors: Daniel Yee1, MD; Nathan Wong1, MD; Sarah Hochendoner, MD; Bradley Collins, MD; Samir Shah, MD. 1These authors contributed equally to this work, The Miriam Hospital, Warren Alpert SOM at Brown University

Introduction: Acquired angioedema due to deficiency of C1 esterase inhibitor (C1INH-AAE) is a rare syndrome of recurrent episodes of angioedema associated with B cell lymphoproliferative disorders and autoimmunity.1 Angioedema is typically self-limiting and affects the skin and mucosal tissues of the upper respiratory and gastrointestinal tracts.2 Here, we present a case where diagnosis of C1INH-AAE was delayed due to under recognition of this rare complication in a patient with chronic lymphocytic leukemia (CLL) and associated autoimmune hemolytic anemia (AIHA).

Case Presentation: Our patient is a 58 year-old Portuguese-speaking female with a history of CLL and AIHA who presented with severe epigastric pain, nausea, and vomiting. She presented with similar symptoms requiring two prior admissions within the last 6 weeks. CT scan of the abdomen and pelvis showed bowel wall thickening and hyperenhancement of the jejunum with mesenteric engorgement. Stool Carey-Blair and Clostridium difficile PCR testing were negative; her symptoms improved over one to two days, and were attributed to viral gastroenteritis. She presented four weeks later with similar symptoms. Repeat CT scan showed persistent, but improved inflammatory change involving the jejunum. She was again diagnosed with viral gastroenteritis, treated with supportive care and had subsequent improvement of symptoms within 24 hours. A hepatobiliary iminodiacetic acid scan was normal and outpatient colonoscopy was arranged.

Four days after being discharged, she presented once again with similar symptoms. A small bowel follow through during this admission was normal. Upon review of her medical history, her recurrent abdominal pain, nausea, and vomiting began shortly after completing a prednisone taper to treat AIHA related to her CLL (diagnosed nearly six months prior to this presentation). She had been started on prednisone 90 mg daily for one week, then tapered to 20 mg daily and weaned off steroid therapy three weeks before her first hospitalization for these recurrent symptoms. Our differential at this time included C1INH-AAE as a complication from CLL vs. small bowel vasculitis vs. inflammatory bowel disease. Laboratory results showed very low C4 (<3), normal C3 (102), normal C1q binding (<1.2), and very low functional C1 esterase inhibitor (15%). She was recently started on Rituximab for treatment.

Discussion: This case illustrates the difficulty in recognizing acquired small bowel angioedema secondary to C1 esterase inhibitor deficiency. Given her history of CLL and recent episode of AIHA, our patient had two underlying, albeit concomitant, risk factors for C1INH-AAE.3 She required recurrent hospital admissions with extensive workup, including multiple laboratory studies and imaging series contributing significantly to healthcare expenditures, before the diagnosis was elucidated. Consideration of this potential differential is key in accurately working up the patient with recurrent, unexplained abdominal pain, diarrhea, and nausea/vomiting with history of lymphoproliferative or autoimmune phenomena, and especially in patients with both conditions.

References


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CALIFORNIA CLINICAL VIGNETTE POSTER FINALIST - SOHAIL YOUSUFI, MD

A Rare Cause of Sudden Cardiac Arrest in a Young Male: Anomalous Left Coronary Artery originating from Pulmonary Artery (ALCAPA)

Authors: Sohail Yousufi MD, Christopher Woo MD, Jessica Murphy DO

Introduction: Anomalous Left Coronary Artery originating from Pulmonary Artery (ALCAPA), also known as Bland-White-Garland syndrome, is a rare congenital condition that is most commonly identified in infants and children. [1] However, it is increasingly being identified in teens and adults. [2] We present one such case of a 20 year old male who had sudden cardiac arrest (SCA) after strenuous exercise and was found to have ALCAPA. The diagnosis was made using transthoracic echocardiography (TTE), coronary CT angiography (CCTA), and coronary angiography. He underwent a successful surgical repair to relocate the left coronary artery to the aorta.

Case Presentation: A 20 year old male collapsed after running on a treadmill in a gym. Bystanders reported that he was unconscious and pulseless. An AED was applied and a shock was delivered, which resulted in return of spontaneous circulation. He was transferred to a hospital, cooled according to a hypothermic protocol and had complete neurologic recovery within 2 days. EKG was notable for left axis deviation and left ventricular hypertrophy (LVH); there was no evidence of arrhythmia, accessory pathway, or Brugada pattern. Interestingly, TTE did not reveal LVH, instead noting that the left ventricle had normal size, wall thickness, and ejection fraction; thereby ruling out hypertrophic cardiomyopathy. However, TTE did identify abnormal Doppler flow between the left coronary artery (LCA) and the pulmonary artery (PA). Also notable was mild mitral regurgitation (MR). There was no EKG, laboratory, or echocardiographic evidence of acute coronary syndrome. CCTA was suggestive of the LCA arising from the main PA, which was further corroborated by invasive coronary angiography. This also identified a complex coronary venous fistula involving the RCA, the left anterior descending (LAD) artery, and the left circumflex artery. Operative findings confirmed that the LCA orifice was located on the posterior side of the pulmonary trunk. Additionally, the RCA and the PA both appeared dilated, and there was substantial collateralization with the LAD. The patient’s ALCAPA was successfully repaired in an open-heart surgery by grafting the LCA onto the ascending aorta and patching the main PA.

Discussion: Typically, ALCAPA presents as heart failure in infancy and childhood due to myocardial ischemia. The likely factors that allowed for this patient’s delayed presentation of ALCAPA are the dominant right coronary artery system and the development of substantial collaterals with the left coronary artery system. Aside from common post-surgical complications of CABG, long term management will entail monitoring for sequelae of long-standing subclinical left-sided cardiac ischemia. This includes monitoring his MR, which may be secondary to papillary muscle ischemia, evaluating for occult malignant arrhythmias, and monitoring for left-sided heart failure. [3] The tortuosity and dilation of his RCA may predispose the vessel to thrombosis or aneurysm. Given his SCA, an ICD may also provide mortality benefit. [4]

References

Beyond Eye Opening: A Case of Non-Cirrhotic Portal Hypertension from Hypervitaminosis A

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Introduction: Chronic hypervitaminosis A can occur from excessive doses over the course of months to years. In high doses, vitamin A is a direct hepatotoxin that accumulates in stellate cells, leading to liver injury and even frank cirrhosis. We describe an unusual case of portal hypertension complicating chronic vitamin A overuse.

Case Presentation: A 46 year old male presented with a one week history of abdominal pain associated with early satiety, diarrhea, and lower extremity edema. The patient had not seen a physician for over 25 years and was only taking vitamin supplements. He drank vodka socially and denied drug use. Physical exam was remarkable for alopecia, a positive fluid wave, and lower extremity edema. Labs were notable for: platelets 106K, Na 128, AST 87, ALT 50, INR 1.2. Ultrasound demonstrated a smooth liver with possible fatty infiltration and large ascites. Paracentesis yielded 1.5L straw-colored fluid. SAAG was 2.8g/dL with total protein of 1.5g/dl; cultures and cytology were negative. Evaluation for autoimmune, metabolic, and infectious causes of hepatitis was negative. Transjugular liver biopsy showed an elevated hepatic venous pressure gradient of 15mmHg. On histology, there was mild chronic hepatitis with perisinusoidal fibrosis and an increased number of lipid-laden stellate cells. Serum retinol level was 195µg/dL (30-72.) On further questioning, the patient revealed that he had been taking 50,000 IU vitamin A for the past year as he heard it was good for his vision. He made a full recovery after discontinuing vitamin A.

Discussion: This case illustrates the importance of a thorough evaluation of patient supplement use, especially considering that over 50 percent of Americans report taking dietary supplements. Acute toxicity can occur with a single dose of >660,000IU of vitamin A and patients often present with nausea, vomiting, vertigo, and blurry vision. More often, vitamin A toxicity presents from chronic daily ingestion of around 50,000 IU of synthetic vitamin A over months; toxicity can be exacerbated by underlying liver and kidney dysfunction, alcoholism, and certain medications. Patients may present with liver disease, alopecia, ataxia, joint pains, and visual impairment. Serum vitamin A levels are not always elevated but liver biopsy showing lipid-laden stellate cells with variable sinusoidal fibrosis is diagnostic; vitamin A is stored in the stellate cells of the liver. Recognition of this condition and appropriate treatment often leads to gradual resolution of symptoms, though there are reported cases of vitamin A-induced liver failure requiring transplantation.

The importance of a detailed history of non-prescription supplements, herbal preparations, and vitamins cannot be overemphasized. This case illustrates a serious, potentially fatal consequence of excessive vitamin A use and is an important reminder to physicians to query patients about the “hidden” medication list.

References

Sexually Transmitted Renal Failure: “A Case of Membranous Glomerulopathy in the setting of Secondary Syphilis”

Authors: Deborah Akanya, MD, Prathiba Phuyal, MD, Pramod Phuyal, MD, Dipesh Patel, MD, Daniel Afrifa, MD and David Regelmann, MD.

Introduction: Membranous glomerulopathy (MGN) is a cause of renal failure that occurs by itself or in conjunction with several other diseases like syphilis. Here we describe a case of MGN in the setting of secondary syphilis (SS).

Case Presentation: A 47-year-old-female sex worker with a history of hepatitis C (HC) with undetected viral load 2 years prior, heroin and cocaine intravenous drug use who presented to the emergency room for evaluation of diffuse rash, abdominal pain, nausea, vomiting, and non-bloody diarrhea, that began one week prior. Her vitals were unremarkable. Her rash was non-tender, well-defined, circular, copper-colored, and macular-papular without necrosis or purulence. CBC showed hemoglobin of 7.8gm/dl, erythrocyte sedimentation rate of 62, C reactive protein of 6.6 mg/L. BUN was 106mg/dl, creatinine (Cr) was 10.9mg/dl, bicarbonate was 10mmol/L, potassium was 5.2 mmol/L, phosphorous was 6.1mg/dl, with estimated GFR of 4 mL/min/1.73 m2. Serum albumin was 2.7gm/dL and cholesterol was 187mg/dL. Urinalysis showed protein > 1000 and microscopic hematuria with urine protein/Cr ratio of 22. Urine toxicology was positive for opioids and cocaine. Renal ultrasound showed bilateral lobulated, enlarged, bulky hypechoic kidneys without hydronephrosis. Cr trended up to 11.7mg/dl despite IV fluids, and she was evaluated by nephrology for emergent hemodialysis (HD). Rapid plasma reagin was reactive, confirmed by Treponema pallidum Ab by TP-PA. HC viral RNA was detected with negative cryoglobulins. Serologies for hepatitis B, HIV, ANA, MPO/PR3, ANCA, and antistreptolysin O were negative. Complement levels were normal. Serum protein electrophoresis showed an M-spike. Renal biopsy revealed stage 2 MGN. Electron microscopy showed one hundred percent foot process effacement with global subepithelial electron dense deposits. Serum PLA2R was negative. She was treated with benzathine penicillin with improvement in the rash, and downtrend in Cr to 3.6, HD was discontinued after 3 sessions, she was counseled on safe sexual practices and discharged with close follow up.

Discussion: Identification of a secondary cause of MGN is uncommon. As of 2013, the incidence was 5.3 cases/100,000. The exact mechanism for syphilis triggering immune complex formation and MGN is unknown. These complexes deposit beneath podocytes on the subepithelial surface of the glomerular basement membrane, resulting in podocyte detachment and increased glomerular permeability to large molecules. Only 5 cases of MGN secondary to syphilis have been described in the last 20 years. These cases involved men with high-risk sexual behaviors and co-infection with HIV or hepatitis. Clinicians must consider secondary causes of MGN as treatment of the underlying condition aids in simplifying treatment. In conclusion, syphilis is a re-emerging infection in the western world, and clinicians should have a high index of suspicion for this cause of MGN in patients presenting with elevated Cr, typical rash and consistent risk factors.

References
Rare Complications of Granulomatosis with Polyangiitis (GPA)

Authors: JAISON JOSHUA, M.D, NAVYA KUCHIPUDI, M.D

Introduction: Granulomatosis with Polyangiitis (GPA, previously known as Wegener’s Granulomatosis) is a rare form of systemic vasculitis with three pathological hallmarks: granulomas and inflammation, necrosis of tissues, vasculitis. Annual incidence is approximately 10 per million. GPA affects small to medium sized vessels with involvement of upper (sinus, nose, trachea) and lower airways, glomerulonephritis and positive PR3-ANCA. It presents sub-acutely and has a strong predilection for white population, particularly among descendants of northern European ancestry. Gender predilection for males to females is 1 to 1. Mean age at diagnosis is 50, less commonly seen in children.

Case Presentation:

CASE#1
Patient is a 74-year-old Caucasian female with left lung collapse secondary to left main bronchial stenosis and necrosis. Serology showed an elevated c-ANCA of 1:320. She had evidence of prior renal disease with trace proteinuria and hematuria, but no active sediment.

CASE#2
Patient is a 78-year-old Caucasian female who presented initially with bilateral hearing loss, ear fullness and constitutional symptoms. She then developed right-sided otomastoiditis and subsequent facial nerve palsy, which was presumed to be secondary to local compression/destruction while traversing in the middle ear.

CASE#3
Patient is a 28-year-old Caucasian male with renal failure, lung abscess, recurrent sinusitis, tracheitis found to have extensive destruction of his nasal septum, nasal turbinates as well as scleritis, which had progressed to scleromalacia perforans.

Discussion: In this case series we present three different patients with three different serious and rare manifestations of Granulomatosis with Polyangiitis (GPA). Patient described in Case#1 had near complete bronchial stenosis of left main bronchus. Treatment options include bronchoscopy-guided dilatation, stenting, laser therapy among others, but unfortunately have high failure rate. Second patient suffered from facial nerve palsy, which occurs only in about 5% of patients with GPA. It is likely secondary to local compression/destruction of facial nerve from otomastoiditis and osteomyelitis. Third patient developed Scleromalacia Perforans, which is a severe necrotizing anterior scleritis that is painless due to necrosis of pain fibers but can lead to spontaneous perforation of globe, if left untreated. These findings demonstrate extensive destructive nature of GPA if left untreated leading to severe comorbidities, poor quality of life and even death warranting early and aggressive treatment.

References

A case of primary peritonitis following a colonoscopy: cause or coincidence?

Authors: Kavya Kelagere Mayigegowda MD, Rinad Tabbalat MD, Shazia Samanani MD, Kostas Papamarkakis MD

Introduction: Colonoscopy is a relatively low risk procedure used for screening, diagnosis and treatment of diseases of the colon. Peritonitis is an infection of the peritoneum and is deemed spontaneous or primary if it occurs without any intra-abdominal source of infection. Primary peritonitis is a rare complication of colonoscopy and related procedures such as polypectomy. We present a case of primary peritonitis following a colonoscopy in a patient with metastatic colon cancer.

Case Presentation: A 20-year-old male with a family history of Hepatoblastoma in his brother, presented with a two-week history of intermittent left lower quadrant abdominal pain. He was afebrile and initial work-up showed a normal White blood cell count (WBC), Hemoglobin (Hg) 7.1 g/dl, Mean corpuscular volume (MCV) 71 femtoliters and a positive stool occult test. A CT scan of the abdomen showed diffuse thickening of the left colon wall, streaky nodular stranding of the omental fat and mild ascites. A CT scan of the chest showed multiple pulmonary nodules. Colonoscopy revealed hundreds of non-bleeding polyps concerning for familial adenomatous polyposis (FAP) with biopsy results showing adenomatous polyps. Subsequent biopsy of a lung nodule showed metastatic adenocarcinoma of colonic origin. Two days after the colonoscopy, he developed worsening abdominal pain, and a distended rigid abdomen on an exam. Repeat CT scan revealed new large volume loculated ascites as well as peritoneal thickening consistent with peritonitis. There was no evidence of intestinal perforation on CT scan. Ascitic fluid analysis from paracentesis revealed 11,000 WBCs and cultures grew group C streptococci. He was started on broad-spectrum parenteral antibiotics with evidence of complete resolution of the collection on repeat CT scan. However, one week later, the patient returned with a fever and recurrent abscess on CT scan, which was not amenable to percutaneous drainage. He underwent surgical drainage of the abscess. He recovered from surgery well and was started on chemotherapy pending genetic testing for FAP.

Discussion: To our knowledge this is the first reported case of primary peritonitis following colonoscopy in a patient with malignant ascites. Malignancy is an overall immunocompromised state, which poses a risk for infections including peritonitis. Although causality cannot be established in our case, we display a temporal sequence of association between the colonoscopy and the onset of peritonitis. Theoretically colonoscopy and related procedures increase the transmural migration of normal flora from the bowel into the peritoneal cavity, which can lead to peritonitis. However, due to lack of sufficient evidence prophylactic antibiotics are not recommended in patients undergoing colonoscopy. Physicians should be cognizant of this potential complication following colonoscopy, especially in patients who are vulnerable to infections such as those with malignancy.

References

Gabapentin Withdrawal presenting as catatonia and altered mental status

Authors: Aziz Khan, Mariam Saeed, Anuradha Kapur

Introduction: Gabapentin is one of the commonly prescribed medications for a variety of medical conditions. Gabapentin is generally well tolerated and withdrawal symptoms like agitation, anxiety, irritability, diaphoresis and tachycardia have been reported in patients with abrupt discontinuation. Catatonia and altered mental status is a rare complication of Gabapentin withdrawal.

Case Presentation: A 77-year-old lady with past medical history of congestive heart failure, hypothyroidism, Hypertension, Obesity, Obstructive sleep apnea and depression was transferred to Saint Francis Hospital (SFH) with altered mental status. Around 8 weeks prior to presentation she had underwent C-spine surgery for C5-C6 spinal fusion and was discharged to long term care physical therapy facility. She was discharged home from the facility after spending 5 weeks. At baseline she was independent in activities of daily living and was alert and oriented upon discharge from the facility. Three days after discharge she was found to have altered mental status and was treated at an outside hospital for Urinary tract infection with ceftriaxone for 7 days. She stayed there for 2 weeks but her condition did not improve so she was transferred to Saint Francis Hospital for further management. On admission she had normal vital signs. She was awake but had a blank stare, was unable to communicate and was not following any commands. She had some rigidity in extremities but was moving all four limbs. Initial work up was negative for infection/sepsis and she had normal urea and electrolytes. She had TSH of 8.13 and fT4 0.9. MRI head showed minimal chronic ischemic white matter disease and was negative acute cerebrovascular event. MRI cervical spine showed central canal stenosis of lower cervical spine and there was no definite abscess. EEG was negative for seizures but showed encephalopathy. CSF analysis was negative for Meningoencephalitis. It was then discovered that she was previously on Gabapentin 1200 mg TID which was discontinued since discharge from the facility. She was then started on, hospital stay day 6, Gabapentin 500 mg BID and 24 hours later she started talking and was back to her baseline mental status.

Discussion: Catatonia following withdrawal from GABA-ergic drugs has been reported in literature. Most of these cases have been reported in the setting of alcohol1 or benzodiazepine2 withdrawal. Zolpidem withdrawal catatonia is also reported3. There have been one other report of catatonia following abrupt taper of Gabapentin in a patient with a bipolar disorder 4.

This case illustrates development of catatonia and change in mental status following abrupt discontinuation of Gabapentin, which resolved with re-initiation of the medication. To our knowledge this is the second such case reported in literature. Gabapentin discontinuation if desired should be done through a slow taper and re-introduction of Gabapentin appears the appropriate treatment in case of withdrawal symptoms.

References

Alteplase Induced Pituitary Apoplexy: A No Brainer.

Authors: Oriana Ramirez Alvarado MD1, Daniel Reef MS-IV2, Yury Malyshev MD1, Oluwo Oluwaseun MD1, Sonu Sahni MD1, 1) Department of Internal Medicine, Brookdale University Hospital Medical Center. Brooklyn, NY, 2) New York Medical College, Valhalla, NY

Introduction: Alteplase is a tissue plasminogen activator (tPA) indicated for the treatment of acute ischemic stroke (AIS)(1). Serious known adverse reactions include hemorrhage and angioedema (1,3). The use of alteplase in AIS is dictated by complex guidelines to avoid adverse events and to offer thrombolytic therapy to patients with a favorable risk to benefit ratio (3). The most common adverse reaction is intracranial, retroperitoneal or gastrointestinal hemorrhage with minor bleeding involving the skin and mucous membranes (1). Herein we describe a unique case of alteplase-induced pituitary apoplexy in the setting of AIS.

Case Presentation: A 68 year old female with HTN, CAD and hypothyroidism S/P partial thyroidectomy presented to the emergency department 30 minutes after sudden onset of slurred speech and weakness of the left face and left arm. Blood pressure was 196/153 and the heart rate was 58 bpm. Physical examination showed decreased sensation of the left face and left arm, left nasolabial fold flattening and left pronator drift. Initial NIHSS score was 5. CT head ruled out intracranial hemorrhage and showed a large sellar mass. Right subcortical ischemic stroke was suspected and tPA was administered 58 minutes after the onset of symptoms. Within 30 minutes patient’s speech improved, NIHSS score decreased to 3 and patient was admitted to the ICU. Post-tPA CT head showed a hemorrhage in the pituitary gland. MRI showed a macroadenoma measuring 2.3x2.5x2.6 cm with internal hemorrhage and suspected evolving pituitary apoplexy. MRA head and neck were negative. Ophthalmic exam showed bilateral inferior optic edema and papilledema. Blood work revealed low FSH/LH (0.9/<0.2 mIU/ml), low IGF (2.7 mg/l), low T4 (0.43 ng/dL), inappropriately low cortisol (5.05 ug/dL) and ACTH (19 pg/ml), borderline normal TSH (1.22 mIU/L - patient on thyroxine), and high prolactin (1243.8 ng/mL). Patient was eventually discharged on cabergoline, levothyroxine and prednisone. Follow up with ophthalmology and neurology revealed, right inferior temporal and left rim visual field defects, unsteady gait, continuous headaches and hand tremors.

Discussion: To the best of our knowledge we describe the first case of alteplase induced pituitary apoplexy. According to the 2018 AHA/ASA guidelines IV alteplase should be administered as soon as possible if there is no intra-axial intracranial neoplasm. Alteplase is still recommended in extra-axial neoplasms (i.e. pituitary masses), but recommendation is based on sporadic reports and expert opinion, not trials. It is not recommended to screen for cerebral microbleeds, but other intracranial structures are not mentioned (3). Our patient shows an example of where screening for microbleeds in extra-axial neoplasms in the setting of AIS is warranted as pituitary apoplexy may complicate administration of alteplase, which adds to morbidity and possibly mortality of AIS patients.

References

Atrial Flutter and Respiratory Distress Following Exposure to Palytoxin

Authors: Valien Kondos, DO, Ahmed Mostafa, MD, David Vearrier, MD, MPH

Introduction: Palytoxin and its analogues are among the most potent toxins known to man, affecting multiple organ systems via oral, dermal, and inhalational routes. While palytoxin has previously been associated with cardiac dysfunction and electrocardiogram (EKG) changes, we report what we believe to be the first case of atrial flutter caused by exposure to palytoxin.

Case Presentation: A 43-year-old male with no past medical history presented with shortness of breath, palpitations, and diaphoresis. He reports aggressively scrubbing his fish tank 3 days prior to presentation, which contained ‘Green Button Polyp’ coral. Resulting particles were aerosolized, inhaled, and there was significant dermal exposure through the arms. Symptoms progressed to metallic taste in his mouth, chills, dizziness, nausea, and cough productive of maroon–colored sputum. He sought medical attention after symptoms proved persistent and progressive. Vital signs on arrival were a temperature of 36.8 ºC, a heart rate of 180-200, a respiratory rate of 54, and oxygen saturation of 88% on room air. Abnormalities on the physical examination included diaphoresis, intercostal retractions, expiratory wheezing, crackles, and an erythematous macular rash on trunk and extremities. Blood investigations revealed an elevated procalcitonin 6.24ng/ml, leukocytosis of 29.9 with 31.1% bandemia, and hyponatremia to 132 mEq/L. EKG revealed narrow complex tachycardia, when temporarily slowed by adenosine revealed 2:1 atrial flutter. Administration of a digoxin dose of 500 mcg, and a total diltiazem dose of 50 mg followed by a 15 mg/hr infusion had no effect on the patient’s tachyarrythmia. The rhythm converted to sinus only after receiving metoprolol 5 mg intravenously. Computed tomography of the chest showed bilateral panlobular ground-glass patchy opacities with intralobular septal thickening. Patient required cardiac intensive care admission and initial respiratory support with non-invasive positive pressure ventilation in the form of BiPAP set at pressures of 16/10 cm H2O at 70% FiO2 which was weaned over the course of 3 days.

Discussion: Palytoxin and its analogues have shown to be present in dinoflagellates of the genus Ostreopsis, which are widely distributed marine environments. The main mechanism of action by which it exerts its effects is by converting the Na/K-ATPase pump into an ionic channel leading to uncontrolled membrane depolarization. Mortality has only been reported after exposure through the oral route, but myriad symptoms have been described following other forms of exposure. Despite being exposed through non-oral routes, this case, however, describes a critically ill patient whose course of illness could have been more complicated had he not presented to care, resulting in further morbidity and even death. Mechanisms of toxicity are not entirely understood and management is supportive. Continued research with updated risk assessment studies appear to be warranted, in addition to increasing physician awareness to aid with diagnosis and patient education.

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Bug Bite or Blood Transfusion? A Fatal Case of West Nile Neuroinvasive Disease

Authors: Michaela A. Seigo, Elizabeth M. Pines, Kristen Facciolo, Michael Benninghoff

Introduction: West Nile virus (WNV) is an RNA flavivirus which can manifest as asymptomatic infection (80%), nonspecific febrile illness (West Nile fever, 20%), or neuroinvasive disease (<1%). West Nile neuroinvasive disease (WNND), which encompasses meningitis, encephalitis, and acute flaccid paralysis, is more common in advanced age, male sex, malignancy, and immunosuppression.

Case Presentation: A 68-year-old man with history of hypertension presented to the Emergency Department in July with subjective fevers, weight loss, and generalized fatigue. Lab work was notable for a hemoglobin of 6.0 g/dL, platelets of 82,000/µL, and white blood cell count of 592,000/µL with 97% lymphocytes. Initial concern was for chronic lymphocytic leukemia (CLL) versus mantle cell lymphoma. The patient received two units of packed red blood cells on hospital day 0 and was started on rituximab on day 2 for leukoreduction. On day 12, the patient began to exhibit lethargy and altered mental status. Due to this rapid decline, he was treated empirically for CLL with bendamustine and rituximab. CT head and MRI brain were unrevealing. On day 17, fluorescence in-situ hybridization confirmed the diagnosis of CLL. Cerebrospinal fluid analysis subsequently revealed a lymphocytic pleocytosis, elevated protein, and positive West Nile virus IgM antibody. The patient required admission to the intensive care unit for encephalopathy, followed by emergent intubation for airway protection on day 20. Unfortunately, he developed septic shock secondary to MRSA pneumonia and bacteremia. He was treated with intravenous immunoglobulin (IVIG) and interferon alfa-2b but ultimately expired on day 31.

Discussion: As of November 13, 2018, over 2,300 cases of WNV have been reported to the Centers for Disease Control and Prevention. After an incubation period of two to fourteen days, WNV commonly presents as West Nile fever, a syndrome characterized by acute-onset fever, headache, malaise, GI symptoms, and a generalized rash. Though typically transmitted by mosquitoes, WNV can rarely be contracted via blood transfusion, organ donation, or breastfeeding. Given the incubation period of up to fourteen days, we hypothesize our patient was exposed to the virus in the hospital setting. Since it is unlikely this exposure came in the form of a mosquito bite, it is plausible he contracted the virus through the blood transfusion on hospital day 0. While all donated blood is tested for WNV via nucleic acid amplification testing, very low-level viremia may go undetected and could have been especially virulent in this immunosuppressed patient. Fulminant, fatal WNND has previously been described in patients with impaired humoral immunity secondary to rituximab. Treatment of WNV is supportive. Studies of IVIG, interferon alfa-2b, and ribavirin have not demonstrated any clear benefit, but are most often utilized in the treatment of WNND in severely immunocompromised patients.

References

Direct-to-Consumer Genetic Testing: A Case of Unnecessary and False-Positive Testing

Authors: Marshall Hoffman, MD; Clesson Turner, MD; Alyson Krokosky, MS

Introduction: Genetic testing capabilities are expanding at a skyrocketing rate and with these ongoing improvements come increased availability of direct-to-consumer genetic testing. There are currently hundreds of companies offering from-home DNA testing for the purposes of determining ancestry, “wellness” qualities, athletic and other personal traits, individualized medication use, carrier status, and pre-symptomatic testing. Additionally, other online resources offer extended interpretation of raw DNA data. By definition, these tests and interpretation services are offered outside of the traditional medical system but nonetheless create implications for the medical community. While there are valid reasons patients seek direct-to-consumer testing, there are a number of significant concerns regarding this type of testing including high false-positive rate, low clinical value, lack of informed consent, lack of appropriate interpretation of results, inappropriate follow-on testing, and exploitation of personal data. In fact, one small 2018 study found a 40% false-positive rate among raw DNA data interpretation services. Research has shown that many patients do not consider the dangers when ordering these tests.

Case Presentation: This is the case of a healthy 18-year-old woman who, along with many family members, sent in a saliva sample to an online ancestral DNA direct-to-consumer company. With the raw DNA file obtained from this company, she utilized online third-party software which analyzed her data and flagged three potentially disease-causing variants in BRCA2, MLH1, and PSEN1. Her many first-degree relatives were in overall good health. Her maternal grandmother reportedly died in her forties from breast cancer and her maternal great-grandmother may have also had breast cancer. There was no family history of early-onset colon, uterine, or ovarian cancer, nor of early-onset Alzheimer disease. After discussion of benefits and limitations of genetic testing, the patient decided to undergo targeted clinical genetic testing for the previously identified BRCA2, MLH1, and PSEN1 variants. Targeted testing by Sanger analysis revealed the patient did not harbor these variants and that the direct-to-consumer results were false-positives. Further genetic testing was not indicated; instead, it was recommended the patient’s mother seek consultation with a genetics clinic.

Discussion: This case highlights the dangers of direct-to-consumer testing and third-party interpretation services that are often not considered by patients. Although having no clinical recommendation for testing of these genes, this patient underwent direct-to-consumer testing which prompted multiple medical visits and confirmatory clinical testing which was ultimately negative. With the growth of the direct-to-consumer genetic testing market, it is expected that primary care providers will increasingly encounter patients who have undergone or have considered this type of testing. These tests have many disadvantages that should be conveyed in an educated manner to the patient. It is important for primary care physicians to be familiar with the types of tests available and how to counsel patients regarding the benefits, limitations, and dangers of these tests.

References

Alien Hand Syndrome – A Rare Presentation of Stroke

Authors: Kelly Le, PGY1, Greater Baltimore Medical Center

Introduction: Alien hand syndrome (AHS) is characterized by intermittent and involuntary movement of a single limb that is not associated with motor dysfunction. AHS may be the initial presentation of stroke in nondominant parietal lobe, corpus collosum tumor or resection, or neurodegenerative diseases. The rarity of AHS and the significant underlying disease it typically represents makes recognizing this condition critical.

Case Presentation: An 88-year-old right handed man presented to the ED complaining of acute onset of weakness, numbness, and intermittent involuntary movement of his left forearm and hand that began three hours prior to presentation. Head CT revealed no hemorrhage, ischemic event, or mass. All labs were unremarkable except for mildly elevated troponin with normal CK-Mb. EKG showed nonspecific ST wave changes, but the patient was asymptomatic. He was admitted for further evaluation and started on ASA and a statin empirically. One day after admission, the patient’s hand movement improved spontaneously. Brain MRI at that time revealed three areas of acute infarct - in the right temporal lobe, right parietal cortex, and right parietal subcortex, suspicious for embolic stroke. Carotid doppler and MRA were unremarkable. TTE showed LVEF of 50-55% and grade I diastolic dysfunction. TEE showed hypermobile septum, probable PFO, but no thrombus. However, after the procedure the patient developed nonsustained atrial flutter which was previously undiagnosed. A probable history of atrial flutter with thromboembolism was considered the likely etiology of his CVA. The patient was discharged home on apixaban, aspirin, metoprolol, and atorvastatin.

Discussion: AHS is rarely encountered, making it an unlikely syndrome to be considered in a differential diagnosis. However, as this case illustrates, AHS may be the presenting complaint of stroke, as well as other serious underlying brain disorders. By being aware of this condition, stroke and other highly morbid brain disorders can be recognized expeditiously and therefore appropriate treatment initiated promptly.
DISTRICT OF COLUMBIA CLINICAL VIGNETTE POSTER FINALIST – NEGIN RASSEKH, MD

The Unmendable Heart

Authors: Negin Rassekh M.D., Medical Resident, Department of Internal Medicine, Providence Hospital, Washington DC

Case Presentation: A 51-year-old Chinese man presented at a hospital due to weakness and chest pain for several days. His past medical history was insignificant, and his only medication was a Chinese herbal medication that he was taking for his heart. On presentation his vital signs were BP 101/70mmHg, HR 165 and RR 30. On physical exam he had an irregular heart rate, crackles on his chest and cold and clammy extremities. Initial workup revealed atrial fibrillation and elevated troponin. A bedside Echocardiogram showed LVEF 20%. He was taken straight to the catheterization lab where coronary angiography showed clean coronary arteries, however right heart catheterization showed that he was in cardiogenic shock. The patient was transferred to the CICU and started on pressors and inotropes. He continued to be hypotensive with unstable SVTs and was taken back to the catheterization lab the same evening for endomyocardial biopsy and placement of an intra-aortic balloon pump. The biopsy report came back 24h later reading Eosinophilic Myocarditis.

The patient was started on high-dose steroids and rheumatology and hematology consults were asked to see him. Imaging and blood work were done to find the cause of his condition but were unrevealing. He stabilized, and steroid doses were tapered, but on day 11 was found unconscious and in sustained VT. He was resuscitated but continued to have rapid ventricular and supraventricular arrhythmias and unfortunately expired.

Discussion: It was concluded that the cause of his Eosinophilic Myocarditis most likely was hypersensitivity due to the Chinese Herbal medication Kyu Shin. Kyu Shin, or its active ingredient Bufotoxin, is a toad venom that has similar characteristics as Digoxin – a medication that is known to cause eosinophilia. The case is a reminder that although many herbal medications are harmless, some may cause significant harm and even – as in our case – death.
AN UNUSUAL CAUSE OF PANCREATITIS

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Introduction: Neoplastic transformation of the intestinal mucosa occurs more commonly near the ampulla than at any other site in the small intestine. Despite this, primary ampullary tumors are rare, with an incidence of approximately four to six cases per million population and account for only 6 percent of lesions that arise in the periampullary region but are responsible for 20 percent of all tumor-related obstructions of the common bile duct. Pancreatitis as a consequence of this lesion is even more rare and the etiological association is often underestimated.

Case Presentation: A 63-year-old Asian male was admitted to the emergency department due to abrupt episodes of intense epigastric abdominal pain radiating to the left flank and back, for 5 hours, not improving after duplicating his daily dose of antacid. Weight loss, nausea, vomits, jaundice were not reported. Past medical history noteworthy for hypertension, benign prostatic hyperplasia and gastritis.

Physical examination revealed diffuse abdominal pain on light palpation, but it was soft and depressible. Rest of the examination was unrevealing. Laboratory studies showed lipase levels greater than 2,000 U/L, blood urea nitrogen slightly elevated, hematocrit in 44% and leukocytosis with left deviation. A diagnosis of acute pancreatitis was made and treated as such.

An abdomen CT scan depicted important edema of peripancreatic fat associated with free fluid in both parieto-colic grooves, wall thickening at the second portion of the duodenum and nodular thickening of Vater’s ampulla. There was no evidence of bile duct dilation or calculus on an abdominal sonogram nor CT scan. Gastroscopy showed a polyp in the gastric corpus and a deformed duodenal papilla with infiltrating mucosa. Biopsy revealed findings compatible with Vater’s ampulla moderate differentiated adenocarcinoma.

Patient had a torpid clinical evolution, requiring mechanical ventilation for a couple of days and parenteral nutritional support. He is currently in route to obtain surgical treatment.

Discussion: In this middle-aged man with no history of alcohol use or evidence of biliary process and negative results for an autoimmune disease, we took the chance of recognizing this Ampulloma as the cause of his acute pancreatitis.

A rare and underestimated cause of a very common disease made this case delightful and proves the relevance in pursuing the etiology of this condition without just limiting to manage the patient’s symptoms.

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Eastern Equine Encephalitis: Importance of Epidemiology and Testing

Authors: Kruti Yagnik, D.O., Ahmad Mahmoud M.D., Jovanna Bertran-Lopez M.D., Kartikeya Cherabuddi M.D., Anthony Cannella M.D., M.Sc.

Introduction: Encephalitis is defined by the presence of an inflammatory process of the brain and the majority of the infectious causes of encephalitis are viruses. Eastern Equine Encephalitis Virus (EEEV) is a mosquito-borne alphavirus that is endemic in the eastern United States. EEEV infection of humans is associated with >30% mortality rate and is classified as one of the most severe of the arbovirus associated encephalitis. It is a rare illness in humans, with an annual average of seven reported human cases.

Case Presentation: A 68-year old Caucasian woman presented with debilitating and progressive neurological deficits; family described her as being physically weak and confused. Upon presentation to the emergency room, she was found to be tachycardic, tachypneic, and febrile (40°C). She was intubated/ventilated due to obtundation. Meningoencephalitis treatment was initiated with vancomycin, ceftriaxone, ampicillin, and acyclovir, and a lumbar puncture was performed. Initial CSF studies demonstrated elevated leukocyte count 810 (mm³) with 73% neutrophil predominance, yet glucose and protein were normal. MRI demonstrated increased T2 signal in deep gray nuclei. Other CSF studies, Varicella Zoster virus, Epstein-Barr virus, Enterovirus PCR panel, Herpes Simplex virus, and serological testing for arbovirus panel (including West Nile Virus) were all negative. An electroencephalogram showed non-convulsive status epilepticus and patient was on multiple anti-epileptic medications. Patients’ autonomic dysfunction triggered repeat CSF collection and sample was sent to the CDC for rabies testing, which was negative. Due to clinical suspicion, arbovirus serology was repeated which resulted with EEE IgM 1:512 with repeat MRI showing progressive diffuse involvement of cortex, temporal lobes, both frontal, and left occipital lobe. Trial of IVIG was initiated, however the patient never recovered neurologically and thus was discharged to a long-term care facility.

Discussion: Clinicians should consider EEE in patients with encephalitis where other workup has been negative, especially if they either live in the eastern United States or visited there. Other clues for EEE are neutrophil predominant CSF, autonomic dysfunction, and MRI changes of basal ganglia and thalamus. It is important to note that the IgM takes 7-10 days to develop, thus it may be a false negative in the initial serum arbovirus panel and should be repeated. According to the Centers for Disease Control and Prevention, there were five confirmed human cases of EEEV in 2018 (three in Florida, one in Michigan, and one in Georgia). In 2018, the Florida Department of Health reported an increased amount of cases demonstrated by positive samples from 52 horses and 153 sentinel chickens across thirty-three counties. The morbidity and mortality rate are very high for EEE and currently there is no treatment or vaccine available. These cases should be immediately reported to the health department so there are community efforts to reduce mosquito populations.
Kaposiform Hemangioendothelioma of the GI tract: An exception to Occam’s principle in an adult with SBO

Authors: Luis E. Aguirre; Robert A. Ali; Darcy A. Kerr; Mahsa Khanlari; Gilberto Lopes

Introduction: Kaposiform hemangioendothelioma (KHE) is a rare and locally aggressive vascular tumor with histological features resembling Kaposi sarcoma and capillary hemangioma mainly occurring in children and adolescents. Approximately 200 cases have been reported since its original description in 1993, with the vast majority presenting at an early age as raised ill-defined lesions with a red-blue hue mainly involving the skin and soft tissues in the extremities. Cases in adults remain extremely rare.

Case Presentation: A previously healthy 29 year-old obese man presented with progressive abdominal pain for 4 months, new-onset nausea/emesis and signs of obstipation found to be consistent with small bowel volvulus. On admission he was tachycardic, febrile and had a distended abdomen that was diffusely tender to palpation with audible borborygmi. Imaging revealed adjacent fat stranding in the mesentery, multiple enlarged mesenteric lymph nodes, questionable pneumatosis intestinalis and free fluid in the pelvis. The patient underwent exploratory laparotomy and resection of 55 cm of necrotic small bowel followed by enteroenterostomy and anastomosis. Copious volume of hemorrhagic fluid was present within the abdomen prior to evisceration of the small bowel. Microscopic examination revealed KHE involving small intestinal mesentery, muscularis propria and submucosa. His recovery was uneventful and he was discharged after stabilization, opting to manage him expectantly with abdominopelvic imaging and to monitor for development of Kasabach-Merritt phenomenon.

Discussion: Kaposiform hemangioendothelioma (KHE) is a rare, locally aggressive vascular neoplasm with histological features resembling those of Kaposi sarcoma (spindle-shaped endothelial cells and slit-like vascular channels) and capillary hemangioma mainly presenting in children and adolescents. The entity was first described in 1993 when it was found to be associated with thrombocytopenia and consumption coagulopathy aka Kasabach-Merritt phenomenon [KMP]. Cases in adults remain extremely rare with 2 reports of lesions involving the testes and 2 cases involving the thoracic cavity or cage.

Immunohistochemically, the spindle cells are positive for vascular endothelial markers (CD31, CD34 and ERG) but not for GLUT1 (which is positive in the endothelial cells of infantile hemangioma). Smooth muscle actin (SMA) is focally positive within the tumor mass indicating the presence of pericytes. The slit-like lymphangiomatous areas exhibiting rich lymphatic vessel configuration show positive staining for D2-40 (podoplanin).

KHE is a locally aggressive neoplasm. Up to 70% of patients with KHE develop Kasabach-Merrit phenomenon, the risk of which seems to be highest with large lesions, congenital lesions and with tumors located in the mediastinum and retroperitoneum. Approximately 10% of patients die as a consequence of disease, either due to local growth or KMP.

Awareness of unusual presentations of KHE such as in this case illustrates how timely surgical intervention and a proper histopathological diagnosis may prevent potentially catastrophic consequences, namely thrombocytopenia and severe bleeding diathesis.

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Large Granular Lymphocytic Leukemia in Sjogren’s Disease Treated with Plaquenil

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Introduction: Large granular lymphocytic (LGL) leukemia is a group of rare lymphoproliferative disorders of large granular lymphocytes: cytotoxic T lymphocytes (CTLs) or natural killer (NK) cells. Approximately 40% of patients with LGL leukemia have autoimmune comorbidities. A strong association between LGL leukemia and rheumatoid arthritis has been described, though a less common association with Sjogren’s syndrome has also been reported. Sjogren’s syndrome is a chronic autoimmune disorder characterized by CD4+ Th1 mediated infiltration of exocrine glands. Typically, patients with Sjogren’s present with dryness of the eyes and mouth due to involvement of the lacrimal and salivary glands. Less frequently, patients may also present with extraglandular manifestations such as fatigue, arthritis, and neuropathy. Treatment of Sjogren’s consists mostly of symptom management, complication prevention, and initiation of immunosuppressive therapy in the appropriate clinical setting. Hydroxychloroquine (HCQ) is an immunomodulatory drug that is often used in the management of Sjogren’s, particularly for patients with extraglandular symptoms such as fatigue, arthralgia, or myalgia. Though this drug is regularly used, there is a scarcity of data demonstrating its efficacy. Here we present a case of a patient with Sjogren’s who developed an LGL leukemia that regressed upon initiation of treatment with hydroxychloroquine.

Case Presentation: A 51-year-old woman with a past medical history of type 1 diabetes mellitus presented with symptoms of fatigue, pruritus, severe eye dryness, and xerostomia. Labs were notable for leukocytosis with a lymphocyte predominance, anemia, and thrombocytopenia. She was evaluated by hematology and diagnosed with LGL leukemia based on an increased number of T-LGL cells (16%) on flow cytometry and a T-cell receptor gamma gene rearrangement. As part of her initial work-up for the cytopenias, she was found to have a positive ANA. She was referred to rheumatology for further evaluation. Additional testing was performed, and revealed a positive SSA, salivary protein 1 IgA and IgM, and carbonic anhydrase IgA and IgM antibodies. Her sicca symptoms along with multiple positive antibodies established the diagnosis of Sjogren’s syndrome. HCQ 400 mg daily was started. She had improvement of her fatigue, normalization of her cell counts, and regression of her LGL leukemia without additional treatment.

Discussion: This case describes a patient with a Sjogren’s syndrome-associated LGL leukemia that regressed with hydroxychloroquine. The use of HCQ alone in treating this condition is a novel discovery. It is important for physicians to understand the potential association between Sjogren’s syndrome and LGL leukemia. In a single center, retrospective study analyzing patients with LGL leukemia, 25% of patients with this lymphoproliferative disorder were found to have primary Sjogren’s syndrome, indicating that screening should be performed in these patients. Given the relative safety of hydroxychloroquine, it should be considered a potential treatment for LGL leukemia in the setting of Sjogren’s syndrome.

References


The Pitfalls of Using The 2010 American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) Classification Criteria for Diagnosis of Rheumatoid Arthritis.

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Introduction: In 2010, the American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) established an updated criteria for the classification of rheumatoid arthritis (RA) that are more sensitive and specific. However, these classification criteria have their limitations. Here, we present a case of monoarthritis misdiagnosed and managed as RA based on the updated ACR/EULAR criteria but subsequently disproved by synovectomy.

Case Presentation: A 63-year-old Caucasian male with a history of left hip osteoarthritis presents with 4-year history of pain in the proximal interphalangeal (PIP) joint of the left fourth finger. The pain was 8/10 in severity, worse with movement and associated with morning stiffness. Family history was significant for psoriasis in father. Physical exam revealed swelling and tenderness of the PIP with limited range of motion. X-ray showed periarticular erosions in the PIP with severe fusiform soft tissue swelling. Labs revealed elevated CRP (4.53 mg/L), anti-CCP antibodies (225 u/ml), and normal RF (11.2 IU/mL). The patient was diagnosed with RA, started on Methotrexate (MTX) 15mg/week and received Triamcinolone 10mg intra-articular injection. Anti-CCP was >250 two months later with no clinical improvement. Etanercept 50mg/ml/week was started and MTX dose increased to 20 mg/week. Three months later, anti-CCP remained >250 with no clinical improvement. MRI showed marked inflammatory changes at the PIP. Etanercept was substituted with Adalimumab 40mg/0.8ml/2weeks plus another Triamcinolone intra-articular injection. The patient remained symptomatic. A synovectomy was therefore performed. A sub-centimeter splinter was retrieved and synovial fluid culture grew a rare fungus, Phaeoacremonium sp. MTX and Adalimumab were replaced with Voriconazole 400mg/day. Ultimately, the patient had complete resolution of the synovitis.

Discussion: The diagnosis of early RA can be challenging due to lack of highly sensitive and specific diagnostic criteria. In 2010, the ACR and EULAR developed a newer classification criteria for RA. They investigated factors that can potentially discriminate undifferentiated arthritis based on the likelihood of developing persistent and/or erosive disease which is the new paradigm of RA. Our case did fulfill these criteria with high anti-CCP reactivity but suggests that meeting the ACR/EULAR 2010 criteria should not be conclusive for the diagnosis of early RA. Most patients with chronic inflammatory monoarthritis of >8 weeks’ duration, whose evaluation failed to define an etiology for the arthritis, need a synovial biopsy to rule out an unusual cause such as indolent infection. Also, if suggested and due to their practicality and convenience, high-resolution ultrasonography could be utilized to detect foreign bodies. Our patient had a wooden splinter introduced and an opportunistic fungus, Phaeoacremonium sp. that is found in soil and plants and mainly infects the immunocompromised. This infection can be fatal if disseminated.

In conclusion, although some RA patients can be challenging to diagnose, this case reinforces the need to continually reevaluate patients when they do not respond to what would be an appropriate treatment.

References


Atypical Case of Herpes Zoster Oticus

Authors: Franco Chevalier, MD, David Coradin MD, Joseph Henain MD, Karishma Samtani, MD

Introduction: Herpes Zoster Oticus (HZO) is characterized by vesicular lesions in the external ear and may also affect vestibular nerve function. Atypical cases of HZO can have simultaneous involvement of multiple cranial nerves leading to trigeminal neuralgia, cochleovestibular symptoms, hoarseness, dysphagia, or even tongue deviation. HZO can sometimes be a diagnostic challenge such as in our patient. We present a 66-year-old female who initially presented to her ENT specialist without vesicular lesions but with severe otalgia and vertigo. At that time, she was treated for malignant otitis externa however did not improve. She later presented to the emergency department (ED), was admitted to our inpatient service and diagnosed with HZO.

Case Presentation: A 66-year-old immunocompetent female without significant PMH presented to the ED with complaints of right-sided, progressively worsening throbbing earache of 1-week duration. She endorsed persistent nausea, vomiting, post-nasal drip, and vertigo. The patient rated her pain 10/10 and said it was starting to extend to the right side of the face and neck. She denied hearing loss but endorsed a sensation of ear fullness.

Otologic exam revealed right external ear edema and erythema with multiple grouped vesicles on an erythematous base specifically over the right pinna and anterior wall of the external auditory canal. Ophthalmologic exam was non-contributory. Cranial nerve exam revealed hypersensitivity and facial edema along the distribution of the right ophthalmic and maxillary branches of the trigeminal nerve.

Orbital/Sella CT was obtained in the ED due to the severity and distribution of pain which did not show mastoiditis or middle ear involvement.

Of note, the patient endorsed that 2 days prior to presenting to the ED she was seen by ENT who diagnosed her with malignant otitis externa for which local antibiotic drops were prescribed however were not effective. We admitted the patient and diagnosed her with HZO. ENT was consulted and agreed with diagnosis. She was started on Acyclovir and steroids. Physical therapy was consulted for vestibular therapy after the patient refused medications for vertigo. Ear scrapings from the lesions were sent for PCR which confirmed our diagnosis of varicella zoster virus (VZV). The patient symptomatically improved and was discharged to follow-up with her primary care physician and ENT as an outpatient.

Discussion: In conclusion, HZO should be considered in patients with severe otalgia and symptoms of otitis not relieved by antibiotics. Our patient’s initial presentation was not definitely consistent with malignant otitis externa and an alternate diagnosis may have been considered at that time. HZO can present with otalgia and vestibular symptoms prior to vesicles appearing such as in our patient. Close follow-up is always necessary for such patients to establish the correct diagnosis and provide the correct treatment in a timely manner.

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Acute Saddle Pulmonary Embolism: A Rare Complication of Mycoplasma Pneumonia

Authors: Farrah Chilet, MD; Arshad Muhammad Iqbal, MD; Meryem Maras-Casey, MD; Nikolay Mitzov, MD

Introduction: Mycoplasma pneumonia affects 1% of the population in the United States. It typically affects young individuals. The majority of patients infected with Mycoplasma experience upper respiratory tract infection symptoms, and about 10% of patients infected with Mycoplasma develop pneumonia. Mycoplasma pneumonia has been found to be associated with multiple extrapulmonary manifestations including bulbous myringitis, myalgias, myocarditis, hepatitis and meningoencephalitis; however, a rare complication is pulmonary embolism, which may be life threatening if not diagnosed early and treated promptly. Our case explores the presentation of Mycoplasma pneumonia complicated by acute saddle pulmonary embolism, an association only reported in the form of case reports globally.

Case Presentation: A 75-year-old female without a previous history of personal or family history of Venous Thromboembolism presented to the emergency department with shortness of breath and was diagnosed with Mycoplasma pneumonia. On initial assessment, the patient was found to be in acute hypoxic respiratory failure, unable to maintain oxygen saturation on room air, was started on a venti-mask along with antibiotics. A few days later, the patient began to desaturate while on the venti-mask and required a non-rebreather mask to maintain her oxygen saturation. Repeat chest X-ray showed a possible developing infiltrate on the left side. Antibiotic coverage was escalated to cover gram negative, positive and atypical microorganisms. Telemetry monitoring showed non-sustained episodes of Atrial Fibrillation with Rapid Ventricular Response and Electrocardiogram showed the presence of new onset SIQIIITIII. Computer Tomography Angiography (CTA) of the chest showed acute saddle pulmonary embolism and bilateral lower extremity doppler ultrasounds were negative. Due to the patient's continued desaturation on the Nonrebreather mask, she was upgraded to the Intensive Care Unit, where she was intubated and started on catheter directed thrombolytic therapy to decreased clot burden. Following catheter directed thrombolyis, the patient was continued on antibiotics and progressively improved.

Discussion: The association of acute pulmonary embolism with mycoplasma pneumonia is rare, with only 47 cases reported world-wide, and only one case presenting with a saddle pulmonary embolism diagnosed at autopsy. To our knowledge, this is the first case reported in a live patient with mycoplasma pneumonia complicated by an acute saddle pulmonary embolism. Mycoplasma pneumonia infection is hypothesized to create a prothrombotic state, predisposing patients to developing venous thromboembolism. This entity is important in order to ensure early diagnosis of pulmonary embolism in association with mycoplasma pneumonia and the initiation of early treatment to improve patient outcomes.
A case of Cutaneous Small Vessel Vasculitis (CSVV) associated with Levofloxacin use.

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Introduction: Levofloxacin is a widely used antibiotic with a relatively safe side effect profile in clinical practice. There are previous report of skin hypersensitive reactions described before. These reactions are largely characterized by pruritus, rash, or photosensitivity.

We described a case of an elderly gentleman who developed small vessel vasculitis with cutaneous involvement after a seven day course of levofloxacin for the treatment of an upper respiratory tract infection.

It is important for clinicians to be alerted to this diagnosis if suspected as a drug reaction and proceed with an immediate cessation of the offending agent.

Case Presentation: A 80 year old gentleman with a medical history relevant for gout who presented with new onset acute palpable rash with arthralgias that started less than 24 hours ago while on treatment with oral Levofloxacin for a recent upper respiratory tract infection. He noticed the rash began on the medial part of his feet and quickly ascended up until his buttocks, lower back and lower abdomen with associated bilateral wrist and shoulder pain.

Physical examination showed stable vital signs on admission. His skin examination showed multiple small palpable, non blanching, non tender purpuric lesions of various sizes, mostly seen from his feet up to his lower abdominal and back area. His laboratory workup revealed a CRP of 72.800mg/ml, his ANA screen was positive with a low titer. His C3 was of 84L. Other immunologic markers including complement C4, Cryoglobulins, RF, anti Smith, C-ANCA and P-ANCA were negative. Infectious etiology and renal damage was ruled out. His skin biopsy was performed. His pathology showed some perivascular mixed inflammatory cells. After these results, his remaining levofloxacin was stopped and he was started on prednisone 20mg BID with some improvement of his rash after 48 hours.

Discussion: Cutaneous small vessel vasculitis is an uncommon but potentially serious side effect of quinolones, including levofloxacin. It is characterized histologically by perivascular inflammation with skin and organ involvement signs and symptoms, such as palpable papules affecting most commonly the lower extremities at onset. Associated symptoms include fevers, arthralgias and generalized lymphadenopathy. Most common lab markers include elevated inflammatory markers, most commonly the erythrocyte sedimentation rate.

It is thought to be related to a type III hypersensitivity reaction with deposition of immune complexes and consequent damage to blood vessels by neutrophils.

Diagnosis remain clinical based on the determination of the skin involvement after starting an offending agent. Skin biopsy is recommended when there is a suspicion of other diseases or when symptoms are persistent or worsening.

The main treatment is to stop the offending agent which usually leads to resolution of the signs and symptoms within days to weeks without the need of additional treatment. Most patients with this condition have a benevolent course following drug withdrawal, which should usually allow physicians to avoid overtreating these patients.
Tetrodotoxin toxicit: A rare case of puffer-fish poisoning in Florida

Authors: Jasdip Grewal, MD, Brian Tostan, MD, Sindhura Kompella, MD, Prashant Reddy Yella MD, San Htoo, MD, Spencer Grobois

Introduction: The differential diagnosis for ascending bilateral paralysis include etiologies that are benign to life threatening emergencies. For true acute ascending paralysis, management includes immediate resuscitative measures. Along with these prompt measures, it is crucial to obtain a thorough history to elucidate the etiology as it helps with management. As this case demonstrates, marine envenomation is an under recognized but important cause of morbidity and mortality for patients with this condition.

Case Presentation: The is the case of a 43 year old Haitian American male with PMH of hypertension, chronic kidney disease and social history pertinent for cocaine abuse who had initially presented to the emergency department (ED) with symptoms of nausea and vomiting. He had associated bilateral leg numbness upon first contact with ED staff. Within a matter of hours his symptoms had progressed to bilateral motor weakness of upper and lower extremities with acute confusion. Given impending inability to protect his airway he was intubated and transferred to intensive care unit (ICU) on mechanical ventilation. After more thorough history taking from collateral sources it was reported patient’s grandmother had presented with similar but milder symptoms of weakness, nausea and vomiting. It was discovered the patient and his grandmother had consumed puffer fish earlier in the day just prior to onset of symptoms. His hospital course was complicated with concomitant ARDS, however after appropriate care he was successfully extubated and had complete resolution of neurological symptoms.

Discussion: Puffer fish toxicity is one of many marine envenomation syndromes that can afflict humans. The pathophysiology lies in the tetrodotoxin, a neurotoxin which blocks sodium channels resulting in skeletal muscle paralysis. It is usually contained in the liver and skin of the pufferfish. Onset of symptoms usually occurs within 20 minutes to 3 hours from consumption. Symptoms include paresthesias, nausea, vomiting with abdominal pain, feelings of doom, and weakness. A rapid ascending paralysis can develop in more severe cases with associated bulbar paralysis and death by respiratory failure within 6-24 hours if not addressed. Treatment is largely symptomatic and supportive. Once life-threatening problems have been addressed, removal of unabsorbed toxin may be attempted by induced vomiting or gastric lavage or by giving activated charcoal. Critically-ill patients must be transferred to ICU and vital signs and oxygenation should be carefully monitored. Symptoms resolve in a few days and prognosis is usually good if the patient survives the first 24 hours. Diagnosis is usually based on clinical signs and symptoms and a history of ingestion. There is no specific lab to confirm toxin ingestion or antitoxin. This case highlights how a thorough history is critical in patients who present with acute neurologic deficits.
Tricuspid valve thrombus: A thromboembolism-in-transit

Authors: Elizabeth Hidlebaugh, MD; Dhishna Chaudhary, MD

Introduction: Tricuspid valve thrombus is rare and reported patients often have antiphospholipid antibody syndrome or structural heart disease. We describe a case of tricuspid valve thromboembolism from bilateral deep vein thromboses (DVTs) that resulted in a massive pulmonary embolism (PE) in a patient with history of malignancy.

Case Presentation: A 59-year-old Guyanese-American female with a past medical history of metastatic uterine and ovarian cancer on chemotherapy presented with dyspnea, bilateral lower extremity (BLE) pain and swelling. Patient was tachycardic and hypotensive requiring fluid resuscitation and norepinephrine. ECG showed S1Q3T3 and her proBNP was 933. BLE venous Dopplers revealed right and left common femoral, femoral and popliteal DVTs. Due to renal insufficiency, V/Q scan was obtained showing intermediate probability for PE. Transthoracic echocardiogram (TTE) revealed a large thrombus attached to the tricuspid valve crossing from the right atrium to the right ventricle. Cardiothoracic (CT) surgery was thus consulted but no thrombectomy was recommended.

Patient was admitted to the ICU. Heparin drip was initiated and IVC filter was placed. She had nearly occlusive thrombus within the left common iliac vein extending into the IVC. Patient was given systemic tPA for 2 hours and heparin drip thereafter. Repeat TTE showed no thrombus and it was suspected that it likely embolized to the pulmonary artery. Patient underwent bilateral LE catheter-directed thrombolytic therapy. She had a CTA of the chest (after her creatinine improved) which revealed massive bilateral PEs. Systemic tPA was given again. Bilateral pulmonary artery catheter-directed thrombolysis was also started but patient went into cardiogenic shock and asystole. Patient was a DNR and expired.

Discussion: Although tricuspid valve thrombus is rare, it is associated with poor prognosis with most deaths occurring in the first 24h of hospitalization. Thus, early diagnosis and treatment are crucial, but no consensus exists on the most effective treatment. In our case, urgent tPA and catheter-directed thrombolysis was used. Though the role of surgical embolectomy has not been well investigated, perhaps early involvement of CT surgery could have decreased the patient’s mortality by improving her clot burden and respiratory symptoms.
Mesenteric Panniculitis: Idiopathic Finding or Paraneoplastic Syndrome?

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Introduction: Follicular lymphoma is the most common of the indolent non-Hodgkin’s lymphomas. It is often widespread at the time of diagnosis, with the gastrointestinal tract being the most common extralymphatic site of involvement.

Mesenteric panniculitis covers a spectrum of disease processes characterized by inflammation and scarring of the adipose tissue of the mesentery; evidence is growing that it may occur as a paraneoplastic syndrome. Histologic findings include chronic inflammation involving the adipose tissue with fat necrosis and fibrosis.

We present a case of a patient diagnosed with disseminated follicular lymphoma who displayed radiographic evidence of mesenteric panniculitis one year before diagnosis. Two years later, after completion of systemic therapy, he exhibits pathologic complete response however again developed findings of mesenteric panniculitis on imaging and pathology.

Case Presentation: A previously-healthy 53 year-old man presented to an emergency department in Miami, FL, complaining of 24 hours of nausea/vomiting and painless abdominal fullness. Since the symptoms began he had not passed stool or flatus. He was diagnosed with small bowel obstruction and treated with conservative management achieving complete resolution of symptoms. At a follow-up appointment one week later, MRI performed to evaluate for intestinal masses revealed findings suspicious for mesenteric panniculitis. Colonoscopy was ordered and was negative for masses. As the symptoms had resolved, no further workup was performed.

One year later, the patient presented to the emergency department complaining of one day of nausea/vomiting and painless abdominal bloating. CT scan showed extensive mesenteric fat stranding with lymphadenopathy more extensive than on previous imaging. Endoscopy revealed duodenal masses, which were biopsied. Workup revealed Stage IV follicular lymphoma; the patient completed first-line therapy with bendamustine plus rituximab. He achieved pathologic complete response however was found to once again have mesenteric panniculitis on imaging and pathology.

Discussion: We present a case of disseminated follicular lymphoma in a patient initially presenting with small bowel obstruction with radiographic evidence of mesenteric panniculitis, successfully treated with conservative management. Etiology of obstruction was not further investigated despite the fact that most small bowel obstructions are known to be caused by either adhesions, hernia, or malignancy. One year later, the symptoms returned and workup revealed disseminated follicular lymphoma. After treatment and achievement of pathologic complete response, he remained with radiographic evidence of mesenteric panniculitis, with PET-positive lesions that contained fat necrosis, histiocytic inflammation, and reactive fibrosis, and negative for lymphoma. Mesenteric panniculitis is an idiopathic condition involving inflammation of mesenteric adipose tissue and recent evidence including several retrospective analyses suggest, with statistical significance, that it may occur as a paraneoplastic syndrome most often associated with gastrointestinal or lymphoproliferative malignancy. Patients with small bowel obstruction and findings on imaging characteristic of mesenteric panniculitis should warrant workup for underlying malignancy if the etiology is ever unclear.
FLORIDA CLINICAL VIGNETTE POSTER FINALIST - TARIQ JABER, MD, MPH

Epidural Abscess secondary to BCG Instillation Therapy for Transitional Cell Carcinoma In Situ

Authors: Tariq Jaber MD MPH, PGY-2; Amy Surti DO MS PGY3

Introduction: Bacille Calmette-Guérin (BCG) is a live-attenuated strain of Mycobacterium bovis that been used for immunization against tuberculosis. It has gained more prominence and has become the mainstay of superficial non-invasive bladder cancer. Treatment is considered safe though adverse reactions do occur. Complications include sepsis, hepatitis, pneumonitis, osteomyelitis, abscess, and prostatic joint infections.

Case Presentation: Here we present a 86 year old male with known osteoporosis; untreated vertebral compression fractures; and transitional cell carcinoma in situ treated with multiple fulgurations and intravesical BCG therapy immunotherapy (3 year duration). He did not report any fevers, chills, night sweats, hematuria, or post-operative complication. He denied having radiation, chemotherapy or other surgical intervention and was in remission at time of presentation. He endorsed progressive weakness of two weeks duration and experienced multiple falls; he required full ambulatory support. He denied incontinence of urine and stool. Physical exam revealed markedly decreased lower extremity muscle strength with decreased reflexes bilaterally, and intact sensation and anal sphincter tone. MRI studies demonstrated subacute compression fractures of T10 and T11, most severe at T10, with a ventral epidural mass causing severe spinal canal stenosis and thoracic spinal cord compression. An emergent decompressive thoracic laminectomy (T9-T11) in addition to having a resection of his ventral epidural mass (T10-T11) was required. Initial pathology report noted a necrotizing granulomatous inflammation with acid fast organism, negative for fungal and carcinoma. Follow-up results was positive for MTB complex. Results for MDDR and Quantiferon gold are pending. A nine-month treatment was initiated with first two months of rifampin, ethambutol and isoniazid; the remaining seven months treatment included rifampin and isoniazid.

Discussion: To date, 22 cases of BCG-related infection have been reported with varying time-frame with respect to BCG treatment and onset of symptoms. Reported risk factors for complications include transurethral resection of the prostate or bladder, trauma from catherization, deep bladder tumor resection, hematuria, bladder outlet obstruction, and radiation to the pelvis. The only notable risk factor here is advanced age. Fulguration therapy for urothelial tumors is considered low risk as it delivers heat current to a targeted area. It minimizes uroepithelial lining breaks and does not pose any immunosuppressive effects. Thus whilst least damaging, it most likely created the necessary conditions that induced the hematogenous spread. It is documented in the literature that breaks in the urogenital epithelium lining is a risk factor for disseminated infection with BCG. As a risk mitigation strategy, BCG doses could be reduced, but long-term outcomes have not been adequately studied. Additionally, to-date, a retrospective analysis of dose-response, dose-duration, risk factors, and development of disseminated BCG have not been properly investigated. Further analysis into treatments with concurrent intravesical BCG therapy is necessary and may help to lessen the occurrence of disseminated BCG infection.

References

4. Sami Obaid, Alexander G. Weil, Ralph Rahme, Cathy Gendron, Daniel Shedid


Wong Variant Dermatomyositis: More Than Skin Deep

Authors: Danielle Jordan Pierson, MD, University of South Florida, Mahad Mohammed, DO, University of South Florida

Introduction: Wong-type dermatomyositis, first identified by Dr. Wong in 1969, is an extremely rare condition described in fewer than 30 cases in the literature. The disease is characterized by cutaneous hyperkeratotic lesions that resemble pityriasis rubra pilaris. It is unclear whether Wong-variant dermatomyositis is associated with an increased incidence of malignancy.

Case Presentation: A 31-year-old previously healthy male presented to the hospital with a several week history of facial swelling and several month history of persistent dry scaly rashes involving his extremities. He was treated by a dermatologist with oral steroids and topical antifungals with no improvement in symptoms. He was employed as a migrant farm worker and had recently traveled to Michigan and Texas before returning to Florida. He endorsed left upper quadrant abdominal pain, vomiting, early satiety, weight loss, weakness, 10 days of subjective fevers, and decreased appetite. On exam, his temperature was 100.4°F and the remainder of his vital signs were normal. He was noted to have periorbital edema, erythematous scaly rashes on the MCPs, DIPs, PIPS, and extensor surfaces of the elbows and knees, desquamation of the palms, and mild erythema of the upper back and chest. He had no appreciable weakness on exam. CPK was normal, LDH was elevated to 572, and ESR was 66. On admission, patient was evaluated by dermatology with concern for amyopathic dermatomyositis. ANA was positive at 1:160 and dermatomyositis antibody panel was negative. Skin biopsies were obtained, which were consistent with the Wong variant of dermatomyositis due to psoriasiform hyperplasia with parakeratosis and interface dermatitis. A CT of the abdomen and pelvis for evaluation of malignancy showed a mass at the body of the pancreas encasing the celiac axis and splenic artery with multiple enlarged lymph nodes. Pathology of the lymph node showed invasive poorly differentiated adenocarcinoma with signet ring cell features, consistent with metastatic gastric adenocarcinoma. An EGD revealed an ulcerated mass with pathology significant for H.pylori, poorly-differentiated adenocarcinoma with signet ring cell features, and erosive chronic non-active gastritis. The patient was treated for H. pylori and started on palliative chemotherapy for metastatic gastric adenocarcinoma with a modified docetaxel, cisplatin, and fluorouracil regimen.

Discussion: Though dermatomyositis is associated with a 5-7 fold increased incidence of malignancy, it is not clear whether this association exists in the Wong variant. This may be due to the young age of previously reported patients or the paucity of data. This case illustrates that all adult patients with suspected dermatomyositis, regardless of the type, should undergo a thorough review of systems to evaluate for any possible underlying etiology for the dermatomyositis as well as an evaluation for malignancy with age-appropriate guidelines.

References

Superior Vena Cava Syndrome in the setting of an Intravascular Lymphoma

Authors: Abdul Khan M.D, Mohammed Faisaluddin M.D, Akriti Jain M.D

Introduction: Intravascular large B cell lymphoma (IVLBCL) is a rare and usually fatal type of extranodal large B cell lymphoma, characterized by selective growth of lymphoma cells within the vascular system. We report a case of a diffuse large B-cell lymphoma (DLBCL) localized in the superior vena cava (SVC) causing an SVC syndrome.

Case Presentation: A 56 year old female presented with complaints of exertional chest pain, dyspnea and swelling over her face and neck. The patient had no significant medical or family history, and her physical exam showed venous distention over her forehead and neck, without any evidence of lymphadenopathy. A CT scan of her chest showed an occlusive mass within the SVC, which was suspected to be an intravascular thrombus. An ultrasound guided thrombolysis was attempted, which was unsuccessful in resolving her symptoms. Subsequently, an MRI showed that the mass was heterogeneously enhancing, raising concerns for an intravascular tumor. Surgical exploration of the mass was undertaken, and it was completely resected from the posterolateral aspect of the superior vena cava. The patient had significant improvement following the surgery, and the mass was diagnosed as a diffuse large B-cell lymphoma on pathologic analysis. It was positive for tumor markers like CD20, CD21, CD23, BCL2, BCL6, MIB-1 and PAX5. FISH analysis did not reveal genetic arrangements involving t(14;18) and MYC, which are reported in high grade lymphomas.

Discussion: IVLBCL is an uncommon and generally fatal disease involving intraluminal proliferation of non-Hodgkin lymphoma cells in blood vessels. In a mediastinal location, it may cause early dyspnea, dry cough and symptoms of superior vena cava syndrome. IVLBCL cells irregularly express several immature and mature B-cell antigens, commonly CD79a (100%), CD20 (96%), MUM1/IRF4 (95%), Bcl-2 (91%), CD19 (85%), immunoglobulin κ light chains (71%), CD5 (38%), Bcl-6 (26%), immunoglobulin λ light chain (18%), CD10 (12%), and CD23 (4%).

Management: Accurate and timely diagnosis of IVBCL is still a problematic issue, due to uncertain clinical presentations and lack of diagnostic protocols. Current guidelines stress the importance of accurate histologic diagnosis prior to starting chemotherapy and the urgent use of endovenous recanalization with stent placement, as necessary. Thrombectomy should be considered prior to stent placement if it is dangerous or difficult to place a stent due to huge thrombi. The tumor mass in our case was surgically amenable and hence was completely resected. Chemotherapeutic options include anthracycline-based regimens such as R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone). The response rates to therapeutic interventions can be satisfactory if initiated at an early stage, with rates of complete remission of about 42%.

Conclusion: IVLBCL can have an aggressive clinical course if left untreated. Its myriad of clinical presentations and lack of pathognomonic features or markers has made antemortem clinical diagnosis difficult and challenging. A timely diagnosis and intervention with combination chemotherapy are important for satisfactory patient outcomes.
A CASE OF RAPIDLY PROGRESSIVE INTERSTITIAL LUNG DISEASE ASSOCIATED WITH MDA-5 POSITIVE DERMATOMYOSITIS

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Introduction: Rapidly progressive interstitial lung disease (RPILD) associated with MDA-5 positive dermatomyositis is a very rare entity; this case demonstrates a particularly devastating presentation and explores the significance of MDA-5 as a biomarker and prognostic indicator.

Case Presentation: A 56 year old female presented to the rheumatology clinic one month after being hospitalized following a fall. During initial hospitalization the patient reported weakness, arthralgias, and generalized rash. Workup revealed elevated inflammatory markers and positive RF. Myositis panel was significant for MDA-5 antibodies but was otherwise negative, including absence of Jo-1 antibodies. This raised suspicion for dermatomyositis associated with RPILD. Baseline PFT’s demonstrated a restrictive breathing pattern and subsequent CT chest showed interstitial opacities and honeycombing of the upper lobes. Patient was scheduled for muscle biopsy and high dose IV steroids but instead insisted on being discharged with oral steroids. Over the ensuing 4 weeks, she developed worsening dyspnea and presents to the clinic now in respiratory distress with worsening weakness and rash. Initial chest radiography was grossly unremarkable and there was no leukocytosis or fever. Bronchoscopy with BAL was preformed in the setting of steroid use in order to rule out infection, and was unremarkable. High dose systemic steroids were initiated but the patient soon progressed into respiratory failure and cardiac arrest within 48 hours of admission. Following resuscitation and mechanically ventilation, CT chest was performed which showed significant interval worsening of interstitial lung opacities consistent with non-specific interstitial pneumonitis. The patient expired later that evening.

Discussion: RPILD is an emerging complication of anti-melanoma differentiation-associated protein 5 (MDA-5) associated dermatomyositis. Patients typically present with erythematous rash with ulcerations and Gottron’s papules. While muscle weakness may be present, an amyopathic variant has shown a higher anti-MDA-5 association. Myositis auto-antibodies including anti-Jo-1 are often negative, thus high suspicion is essential for diagnosis. It is well established in the literature that MDA-5 has a strong specificity for RPILD making it a good diagnostic biomarker. However, muscle biopsy remains gold standard for definitive diagnosis. Recent data has also shown anti-MDA-5 to be a prognostic indicator with its presence correlating with poor prognosis. Therapy for anti-MD-5 associated dermatomyositis includes high dose steroids, along with other immunosuppressants, typically cyclophosphamide, cyclosporine, and sometimes rituxamib. Recent studies have also shown some benefit to plasmapheresis. However, even with optimal treatment prognosis remains extremely poor. Although rare, RPILD should be suspected in MDA-5 positive patients since prognosis is very poor. Survival can sometimes be limited to months, as was this case with this patient who experienced very rapid deterioration. Prompt treatment with proper immunosuppressive therapy may prolong survival and in rare cases cause remission, but is most often ineffective. More research is essential in order to develop more effective strategies to manage this devastating illness.

References

A Rare Disease with Drastic Presentation: Eosinophilic Gastroenteritis presenting as Gastric Outlet Obstruction

Authors: Saritza Mendoza, MD, MHS

Introduction: Eosinophilic gastroenteritis (EGE) is a rare disease with prevalence of 22 per 100,000 persons. EGE can present at any time, but most typically presents in the third and fifth decades with a slight male predominance. The pathogenesis of EGE is not well understood and clinical features of EGE depend on the location, extent, and the layers involved by the eosinophilic infiltration. Patients commonly present with peripheral eosinophilia (average absolute eosinophil count of 1000 cells/µL), elevated serum immunoglobulin E (IgE), and anemia secondary to iron deficiency or occult gastrointestinal bleeding. Eosinophilic mucosal infiltration causes symptoms including abdominal pain, nausea, vomiting, early satiety and diarrhea. Deeper submucosal, muscular infiltration of the gastrointestinal tract results in wall thickening which can impaired motility and present as dysphagia, regurgitation of undigested foods, intestinal perforation and even gastric outlet obstruction. Identification and proper diagnosis for this disease is vital as it can lead to malabsorption, protein-losing enteropathy and failure to thrive.

Case Presentation: A 48 year old female with no past medical history presented with acute onset dysphagia, early satiety, nausea /vomiting and epigastric pain. Eosinophilia noted (absolute eosinophil count was 2,084/µL, reference < 500/µL ). Serum immunoglobulin (IgE) 457 Ku/L (reference <114 Ku/L). CT abdomen revealed marked distention of stomach and a transition point in the first portion of the duodenum. Supportive treatment with IV fluids, nasogastric tube for decompression, and anti-emetics for nausea control was provided. Upper endoscopy was normal. CT enterography consistent with thickening of the distal esophagus and gastric antrum, concerning for a submucosal or serosal inflammatory process. Endoscopic ultrasound with fine needle aspiration (EUS-FNA) showed endosonographic thickening of the submucosal layers of the antrum, pylorus and duodenum. Widespread biopsies revealed diffuse eosinophilic infiltration.

Final diagnosis of this rare disease required collaboration with hematology. Diagnosis was made after extensive workup of eosinophilia to rule out parasitic infections, hematologic malignancies and other immunologic disorders. Due to selective gastrointestinal infiltration, patient was diagnosed with eosinophilic gastroenteritis. Successfully treated with budesonide 9mg daily with strong symptomatic control of the nausea and vomiting, dysphagia and early satiety.

Discussion: Eosinophilic gastroenteritis (EGE) should be suspected in a patient with abdominal pain, nausea, vomiting, early satiety, diarrhea, weight loss, or ascites in conjunction with peripheral eosinophilia (eosinophil count >500 eosinophils/µL in the peripheral blood). Diagnosis is based on eosinophilic infiltration of the gastrointestinal tract on biopsy and/or eosinophilic ascitic fluid, a lack of involvement of any other organs, and the absence of other etiologies for intestinal eosinophilia. Treatment includes a six food elimination diet (avoidance of soy, wheat, egg, milk, peanut/tree nuts and fish/shellfish). Glucocorticoid therapy (prednisone 20-40 mg/day) for acute symptom management and transition to oral budesonide has been used for patients with disease in the gastric antrum. Other therapies for refractory disease include ketotifen (H1 antihistamine, mast cell), montelukast (leukotriene antagonist), and omalizumab anti-IgE monoclonal antibody.
A case of Superior Vena Cava syndrome mimics decompensated heart failure

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Introduction: Pancoast tumor is known to result in superior vena cava (SVC) syndrome, which presents with upper extremity and facial edema. However, lower extremity edema is not a recognized feature. Herein, we describe a case of Pancoast tumor with features of SVC syndrome and bilateral lower extremity edema imitating a worsened heart failure presentation.

Case Presentation: A 77-year-old man with a 30 pack year smoking history and chronic obstructive pulmonary disease presented with one-month of worsening bilateral upper and lower extremity swelling and dyspnea on exertion. He denied chest pain, palpitations, paroxysmal nocturnal dyspnea, or orthopnea. Vitals were normal. Examination showed distended neck veins, but ptosis, miosis or anhydrosis were not noted. He had bilateral upper and lower extremity 1+ pitting edema up to the mid-forearm and mid-calf. He had no calf tenderness or skin color changes.

Initial pro-BNP was 189 and LVEF was 55-65% on 2D Echocardiogram. His duplex ultrasonography of the upper and lower extremities revealed no evidence of deep venous thrombosis. Given his generalized edema, he was initially treated with Furosemide 20 IV twice daily with no clinical improvement. Chest X-ray revealed right sided atelectasis and pleural effusion, right upper lung bullous changes, and increased soft tissue density in the ascending thoracic aorta region of uncertain etiology. This concern for mass led to a computerized tomography with angiography of the chest to be done that revealed a solid lobulated right lung mass (8.4 x 7.2 x 7.2 cm) extending to the hilum, compressing the branches of right pulmonary artery and SVC with opacification of hemiazygous and intercostal veins, along with a right-sided pleural effusion. Apparent filling defects were also present in the left renal vein and inferior vena cava from the occlusion leading to reversal of flow. Chest tube was inserted that revealed chylothorax. Biopsy showed neuroendocrine small cell carcinoma of the lung. The patient initially had good clinical improvement on Cisplatin and Etoposide. However, he started to deteriorate clinically due to medication side effects and developed severe myelosupression, eventually leading to hospice care in the next six months.

Discussion: In addition to upper extremity and facial swelling, compression of SVC by Pancoast tumor can result in reversed blood flow to hemiazygous veins and branches of the inferior vena cava with dilation causing bilateral lower extremity swelling. This case is unique because the patient had lymphedema and chylothorax, suggesting thoracic duct compression leading to filling defects in the setting of small cell carcinoma of the lung. Findings of bilateral upper and lower extremity edema in a patient with significant smoking history should alert the physician to this diagnosis. This could lead to earlier detection of Pancoast tumor, thus resulting in more treatment options and improved outcomes.
A Hypoxemic Puzzle – No Pressure

Authors: Steven Tijmes, DO, Aharon Sareli, MD

Introduction: Right to left intra-cardiac shunts are a common cause of hypoxemia. Rarely, right to left shunts occur, through an ASD, in the absence of significant elevation of right cardiac chamber pressures.

Case Presentation: An 84-year-old female with a history of asthma, osteoporosis, and hypothyroidism presented with chronic progressive shortness of breath and was found to be hypoxemic. As part of her work up, right and left cardiac catheterization was performed two months prior to current presentation. The following parameters were demonstrated: Right atrial pressure of 4mmHg, right ventricular pressure of 26/6 mmHg, pulmonary artery pressure 25/9 mmHg (mean = 15mmHg), pulmonary capillary wedge pressure of 6mmHg, cardiac output of 3.1 l/min, pulmonary artery saturation 74%, superior vena cava (SVC) saturation 73%, arterial saturation 96%. Left heart catheterization revealed no significant coronary artery disease.

Workup
Notable diagnostic results included hemoglobin of 17.3 g/dL and PaO2 of 40 mmHg on room air. CT chest revealed clear lung fields without evidence of acute pulmonary embolism. Echocardiogram with bubble study revealed an atrial septal defect (ASD) with immediate crossing of agitated saline bubbles - causing complete opacification of the left atrium and ventricle. CT coronary angiogram supported evidence of shunting at the atrial level. Right and left cardiac catheterization was performed, including serial chamber pressure and saturation measurement. Agitated saline bubbles were injected in the SVC and inferior vena cava (IVC) with TEE imaging of the atrial septum. Injection into the IVC revealed immediate passage of contrast through the ASD into the left atrium (LA) with dense opacification of the LA. Despite no notable elevation of right-sided pressures, a prominent Eustachian valve in the IVC directed blood flow into the ASD causing right-to-left shunt physiology. The ASD was closed using a 30mm Cardioform Septal Occluder. After placement of the occluder device, repeat bubble study revealed no residual shunt physiology and the patient’s hypoxemia resolved.

Discussion: The rare combination of a right-to-left intra-cardiac shunt in the absence of significantly raised right-sided pressures poses a unique diagnostic puzzle. Rare cases have been described - patients with anomalous IVC valves that direct blood flow into an ASD. This combination may cause right-to-left shunt physiology in the absence of elevated right cardiac chamber pressures (Morishta et al, Bannerjee et al). Similarly, our patient presented with severe hypoxemia, related to shunt physiology through an ASD. Shunting occurred in the absence of significant right sided cardiac pressures. A persistent prominent IVC Eustachian valve was identified as the key factor directing blood flow through the ASD despite any significant elevation of right sided cardiac pressure. It is vital to recognize this possibility as a rare cause of hypoxemia.

References
Fever of unknown origin, altered mental status, diplopia and a retroperitoneal mass as a presentation of Immunoglobulin G4-related disease (IgG4-RD)

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Introduction: Immunoglobulin G4 related-disease is an immune-mediated, fibroinflammatory condition that can affect any organ. It is characterized by lymphoplasmacytic infiltration tissues leading to fibrosis and elevated serum IgG4 levels. IgG4-RD is a relatively novel, unifying clinical entity that is now being diagnosed with increased frequency.

Case Presentation: A 72-year-old gentleman with history of end-stage renal disease and hypertension presented to the ED with history of diplopia for the last 2 days, anorexia and unintentional weight loss. On arrival he was hemodynamically stable and afebrile. Physical exam was abnormal at the eyes exam with inability to abduct the right eye, suggestive of lateral rectus palsy. On the second day of admission, he developed fever that persisted throughout the hospitalization. He also presented intermittent episodes of confusion, severe mood swings, agitation and abnormal behavior. Work up to rule out infectious etiologies included normal blood cell counts, liver function tests and urine analysis. Hepatitis panel, HIV serology, urine and blood cultures were negative. Brain CT was negative for abnormalities. Cerebrospinal fluid tests were normal. Transthoracic echocardiogram was negative for vegetations. Chest CTA and lower extremities ultrasound were negative for thromboembolism. CRP and ESR were significantly elevated. Abdominal CT scan showed a diffuse soft tissue thickening around the kidneys and proximal ureters and a mass-like soft tissue thickening surrounding the distal aorta and aortic bifurcation, extending down to the common iliac artery. These findings were suspicious for retroperitoneal fibrosis. ANA, c-ANCA, p-ANCA, IgA and IgM levels were normal. IgG and IgG4 levels were elevated (2070mg/dl and 280mg/dl). Prior renal biopsy done three years ago was reviewed showing obliterative chronic tubulointerstitial nephritis. The renal cortex had a prominent number of plasma cells with positive immunoreactivity for IgG and IgG4. The IgG4: IgG positive plasma cell ratio was estimated greater than 40%. The patient was diagnosed with IgG4-RD and received a trial of steroids with subsequent resolution of fever and improvement of mental status. At a 2-week follow up in the outpatient clinic, he was found to be afebrile with resolved diplopia, total return of normal mental status and no other complaints.

Discussion: IgG4-RD has a subacute onset and rarely presents with constitutional symptoms like fever, weight loss and elevated inflammatory markers. As we see in this patient, fever of unknown origin has proven to be a significant part of the IgG4-RD spectrum. In our case, the finding of a retroperitoneal mass suspicious for retroperitoneal fibrosis and the results of the kidney biopsy, prompted us to think about IgG4-RD as a unifying diagnosis. Our patient presented multiorgan involvement with systemic inflammatory signs and changes in mental status which are atypical. We report this case in order to increase awareness of an unusual presentation of a relatively novel condition.

References

Cardiac Amyloidosis presenting as a rapid decline in ejection fraction and persistent troponinemia.

Authors: Temidayo Abe, MD., Eric Chang, MD., Bhatia Kapil, MD. Duggal Aarti, MD., Muhammad Bilal. Leonard Gyebi. Associate Professor; Department of Medicine. Morehouse School of Medicine Atlanta GA

Introduction: Cardiac amyloidosis is an important cause of heart failure with a prevalence of 11% in the United Kingdom 1. It remains underdiagnosed and is often misattributed to hypertension despite a high fatality rate. Mean survival is less than 1-year in untreated primary light chain amyloidosis and less than 4-years for wild-type transthyretin amyloidosis 2. We present this unique case to highlight the role of cardiac biomarkers in the early diagnosis of cardiac amyloidosis.

Case Presentation: A 78-year-old African-American male with a history of paroxysmal atrial fibrillation, syncope, hypertension, and heart failure with reduced ejection fraction (HFrEF 30-40%) presented to the emergency department with increased exercise intolerance and shortness of breath. In addition, he reported progressive leg swelling and episodic chest pressure. Vitals were within normal limits (BP 106/71, RR 14, HR 70; T 98F) Physical exam was significant for a non-obese male with +1 JVD, + 1 bilateral lower extremity edema, and inspiratory crackles.

Labs were significant for a troponin (0.27) and BNP (389) that were higher than his baseline (0.14 and 300). Furthermore, persistently elevated troponin (mean 0.145) was noted for approximately 10 years, 5 years before his HF diagnosis. All other labs including kidney and liver function, hematology, and urine drug screen were normal. Chest X-ray revealed no acute cardiopulmonary abnormalities. EKG showed normal sinus rhythm with low voltage but no signs of ischemia. The patient was admitted for management of non-ST elevation myocardial infarction.

Transthoracic echocardiography (ECHO) was obtained which revealed a progressive decline in left ventricular ejection fraction (LVEF) to 25-35% along with worsening diastolic dysfunction with E/A 3.5 and medial E/E’ 24 despite patient’s adherence to medical management. ECHO at HF diagnosis (5 years prior) showed LVEF 50-55% and mild diastolic dysfunction E/A 0.78. Repeat ECHO 1.5 years prior revealed a decline in LVEF to 30-40%. Cardiac MRI was performed to rule out apical ventricular thrombi and showed diffuse late gadolinium enhancement concerning for amyloidosis. Follow up nuclear bone imaging revealed increased cardiac uptake greater than bone, suggestive of grade iii cardiac amyloidosis. Fat pad biopsy and mass spectrometry confirmed tissue transthyretin amyloidosis. The patient was managed conservatively with diuretics and discharged with appropriate follow ups in place. His home lisinopril and metoprolol were discontinued due to concerns that they may worsen amyloidosis symptoms.

Discussion: Persistently elevated cardiac biomarkers may of prognostic value 3. in patients who do not respond to normal HF therapies and have rapid disease progression. Studies have shown that these markers may also be of diagnostic significance 4 and help to differentiate cardiac amyloidosis from other causes of heart failure. In addition, this case illustrates the fact that more frequent ECHOs may be necessary to monitor disease progression in this patient population.

References


Acute onset of delirium and a broken heart from goals of care discussion

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Introduction: As physicians, we frequently engage patients and families in goals of care discussions in the context of advanced serious illnesses. These discussions can be challenging for numerous reasons. We present an atypical case of a woman, who experienced acute onset of confusion, numbness, pre-syncpe, chest pain, and abdominal pain, after initiating a discussion on her husband’s prognosis.

Case Presentation: A 70 y/o female with HTN, fibromyalgia, and anxiety, developed sudden onset of confusion, headache, upper extremity weakness, lightheadedness, substernal and epigastric pain after initiating a discussion on the prognosis of her terminally ill husband. She was sent to the ED for suspicion of having a TIA or CVA. On evaluation, she was afebrile and hemodynamically stable. Her confusion was transient and resolved within an hour. Physical exam was without focal deficits. CT of the head revealed no acute intracranial abnormalities and no evidence of perfusion or flow limiting defects. EKG revealed no abnormalities indicative of ischemia. Serologic workup was remarkable for a troponin that increased from 0.05 ng/mL to 1.57 ng/mL over 6 hours. She was admitted and treated with an NSTEMI protocol. An echocardiogram revealed normal LV size and thickness but a mid-distal septal, apical akinesis without thrombus and an estimated LVEF of 30-35%. The following day, a left heart catheterization revealed mild nonobstructive coronary disease and an abnormal left ventricular wall motion with elevated left ventricular end-diastolic pressure consistent with stress cardiomyopathy. She was stabilized and discharged with metoprolol and grief counseling support.

Discussion: Stress cardiomyopathy is associated with severe emotional or physical stress that can cause rapid weakness of the heart muscle. This case highlights a unique yet rare complication of having goals of care and prognostication discussions with patients’ families. Delivering bad news is very challenging itself, and when compiled with physical manifestations of symptoms, it can be even more so. Our patient developed a constellation of symptoms that manifested from a “broken heart” or stress cardiomyopathy, that after investigation, was attributed to hearing about the terminal state of her husband’s illness. Although its mechanism of myocardial stunning is still unknown, it should remain a possible complication of discussing goals of care or delivering news to loved ones of a patient.
Can Your Emotions Really Break Your Heart?

Authors: Bashar Kadhim, Tanya Reddy, Fasial Fa'ak

Introduction: Spontaneous coronary artery dissection (SCAD) is a rare clinical diagnosis that frequently presents as an acute coronary syndrome (ACS). It occurs secondary to separation in the layers of the arterial wall, creating a false lumen. Hemorrhage into the false lumen can impinge upon the true lumen of the coronary artery, impairing blood flow and causing myocardial ischemia, infarction, sudden death, cardiogenic shock, or pericardial tamponade.

Case Presentation: A 40-year-old healthy female with no ACS risk factors or family history of ACS presented with sudden onset, right-sided chest pain, described as pressure-like. The patient indicated week-long palpitations and dizziness, occurring after several emotionally stressful days in which she learned her sister was comatose secondary to overdose.

The pain started at rest and was associated with shortness of breath. Physical exam revealed HR 67bpm, RR 20cpm, BP 120/94mmHg, temperature 36.6°C, and saturation 100% in room air. EKG revealed no acute ST changes. HEART score was calculated to be 1 (based on positive troponin) correlating with 0.9-1.7% risk of major adverse coronary event; initial troponin was 0.432 ng/ml. Patient was given aspirin, morphine, and started on a heparin drip due to elevated troponin and ongoing chest pain.

The second troponin increased significantly to 3.919ng/ml, and patient was sent immediately to catheterization lab. She was found to have diffuse narrowing (concerning dissection) in the mid to distal LAD with 90% stenosis in the distal LAD. Stent was not placed due to possible progression of dissection proximally. Echocardiogram showed mild hypokinesis of the anterior septum and apical wall with an ejection fraction of 45-55%.

Patient was medically managed with Plavix 75mg, Aspirin 81mg, Metoprolol 12.5mg, and Atorvastatin 80mg. Repeat echocardiography two months later showed no regional wall motion abnormality and an ejection fraction greater than 60%. Stress test showed no ischemic changes.

Discussion: SCAD is a fatal condition occurring predominantly among younger females in the absence of ACS risk factors. In recent studies, 12-48% of patients report extreme stress prior to their diagnosis. Treatment options include medical therapy, percutaneous coronary intervention, or coronary artery bypass graft. A conservative approach is generally favored in stable SCAD and currently is based on an individualized approach/physician experience. Most patients are continued on indefinite antithrombotic therapy with aspirin, clopidogrel, glycoprotein IIb/IIIa inhibitors, which has been extrapolated from known benefits in myocardial infarction. In patients with reduced left ventricular systolic function, β-Blockers and angiotensin-converting enzyme inhibitors (or angiotensin receptor blockers), nitrates, and ranolazine are considered beneficial.

Due to its rarity, physicians should consider SCAD as a differential diagnosis in young, healthy females presenting with typical chest pain and low ACS risk/HEART score. More research is warranted as there are no specific treatment guidelines and much remains to be learned.

References


A False Positive Fourth Generation HIV Test: What Else To Consider?

Authors: Nirja Mehta MD, Brent Gawey BS, Stacie Schmidt MD, Manasi Tannu MD MPH

Introduction: A new diagnosis of HIV can be emotional and life altering. Therefore, every effort should be made to avoid false positive diagnoses. The HIV screening algorithm now employs the fourth-generation HIV test: a combination enzyme-linked immunosorbent assay (ELISA) that tests for both HIV antibodies and the p24 antigen. Despite a reported sensitivity and specificity of greater than 99%, false positive results do occur.1 We present a case of a patient who denied sexual activity or exposure to intravenous needles, who was found to have a positive 4th generation HIV screen.

Case Presentation: A 17-year-old female presented to the Emergency Department (ED) complaining of headache, fever and myalgias. On the of admission, she had also developed a rash on her face and cheeks. During ED evaluation, a 4th generation HIV screen returned positive; she was admitted for treatment of acute HIV infection. Throughout the hospitalization, the patient denied a history of sexual activity or use of needles. Follow-up HIV confirmatory testing and HIV viral load were negative. LP did not reveal any sources of infection. She was then evaluated for rheumatologic causes of cyclic fever, rash and headache. She was found to have +ANA, +dsDNA, +Smith, +RNP, Low C3, Low C4, and elevated anti-Histone. She was diagnosed with systemic lupus erythematosus; symptoms improved with administration of prednisone and hydroxychloroquine.

Discussion: A false-positive HIV test occurs when a positive 4th generation screen is followed by a negative confirmatory test, with either an antibody differentiation assay or HIV-1 RNA qualitative PCR assay.2-4 Given the high sensitivity and specificity of the 4th generation HIV screen, clinicians generally do not search for alternative diagnoses. This case demonstrates the importance of continuing to consider alternative diagnoses while awaiting confirmatory testing. Cases of false positives have been observed in various microorganism infections, pregnancy, conditions with cross-reactive antibodies, vaccinations, and laboratory error. Microorganism infections known to cause false-positive testing include human T-cell lymphotropic virus (HTLV-1/2), Epstein-Barr virus (EBV), viral hepatitis, Mycobacterium tuberculosis, Rickettsia spp., Toxoplasma, and Schistosoma.5-11 Work-up should also consider conditions with cross-reactive antibodies such as current or past pregnancies, collagen-vascular disease, rheumatologic disease, angioimmunoblastic T-cell lymphoma, polyclonal hypergammaglobulinemia, autoimmune hemolytic anemia, and diseases with high levels of anti-nuclear antibody titers such as systemic lupus erythematosus (SLE).10,12 Previous vaccinations with experimental HIV-1 vaccination and influenza vaccination have also been noted to produce false-positive results.13,14 When approaching a patient with a positive HIV screen but low pre-test suspicion, clinicians should consider these alternative diagnoses.

References

A Severe Presentation of Coccidiomycosis in an Immune-Competent Patient

Authors: Abigail Mansch MD and faculty supervisor Rich Doxey MD

Introduction

Coccidioides do not present to medical care because their clinical symptoms are mild. Those that do present may have a wide array of symptoms ranging from cough and fever to cutaneous manifestations. I present a case of severe coccidiomycosis in which the patient developed ARDS and septic shock.

Case Presentation: A 72 year old male with no significant past medical history presented with a productive cough and chills while on a cruise from Vancouver, BC to Alaska. He had no other presenting symptoms. Upon presentation the patient was afebrile and had a leukocytosis. He was initially diagnosed with and treated for community acquired pneumonia. However, he clinically declined and required intubation, ultimately developing ARDS with septic chock requiring vasopressor support. Respiratory samples were obtained, and all of the cultures grew coccidioides. The patient was treated with antifungal medications. After starting the antifungal treatment his hemodynamics stabilized and he was weaned off of the norepinephrine and vasopressin. There was no evidence of fungemia, and lumbar puncture was not consistent with central nervous system involvement.

The patient's history was notable for extensive travel around the world, and he had settled in southern California in the 1990s. Prior to his profound presentation with coccidiomycosis, his only significant medical history was hypertension and hyperlipidemia, and he had no history of being immunocompromised.

Discussion: Coccidioides are an endemic species in the southwestern United States, and most coccidioides infections occur in several counties in Arizona and California. Individuals who are exposed to the arthroconidium of the fungus are susceptible to infection, even in they have lived in the endemic region for many years. However, less than half of all coccidioides infections come to clinical attention because most infections are mild. Additionally, the range of severity of symptoms is broad. Common presenting symptoms include cough, fever, and chest pain. Patients can also develop systemic complaints including arthralgias, cutaneous manifestations, and fatigue. However, severe infection presenting like this patient is less common.

Patients who are healthy and have mild disease do not necessarily require treatment with antifungals, as they typically clear the infection on their own. Patients with severe infection or those at risk of complications (ex: immunocompromised patients, pregnant patients, etc.) should be treated with antifungals. Travel and exposure history can provide critical clues to a patient's diagnosis, even if they have a profound presentation of a typically subclinical infection.

References

A case of hypophysitis induced by immune checkpoint blockade

Authors: Theodora Pappa, Jerry Liu, Chung-Kay Koh

Introduction: The rapid advances in immunotherapy have revolutionized cancer care. Immune checkpoint proteins, such as cytotoxic T-lymphocyte antigen-4 (CTLA4), interfere with immune homeostasis by down regulating T-cell signaling. This prevents uncontrolled T-cell proliferation and maintains tolerance to tumor-associated antigens. CTLA4 blocking antibodies can restore the cytotoxic activity of T-cells towards cancer cells. Systemic administration of the CTLA4-blocking antibody ipilimumab has demonstrated significant survival benefit and improved progression-free survival in patients with advanced cutaneous melanoma. However, immune checkpoint blockade can lead to a syndrome of autoimmune/auto-inflammatory side effects, designated as immune-related adverse events (IRAE), including endocrinopathies. Here we describe a case of hypophysitis induced by the CTLA4-blocking antibody ipilimumab.

Case Presentation: The patient was a 78-year-old female with stage IV melanoma with pulmonary metastases, positive for the BRAF mutation V600K, who received four cycles of treatment with ipilimumab resulting in decrease in the size of the pulmonary metastases. Eight weeks afterwards, the patient reported significant fatigue, diarrhea and blurry vision. Lab work was notable for hyponatremia (Na 120 mEq/l). Hormonal testing was consistent with adrenal insufficiency [cortisol 0.7 ug/dl (normal range: 4.3-22.4), TSH 0.48 U/ml, free T4 1.43 mg/dl, serum osmolarity 281 mosmol/kg, urine osmolarity 549 mosmol/kg, uric acid 3.9 mg/dl]. Brain MRI demonstrated an empty sella. The diagnosis of pituitary insufficiency due to ipilimumab-induced hypophysitis was made and she was started on a methylprednisolone pack with rapid improvement of her symptoms. She was later switched to prednisone 20mg per day with slow tapering and treatment with levothyroxine. On a two year follow up, the patient is receiving substitution therapy with prednisone and levothyroxine. Of note, in the interim she has received therapy with another immune checkpoint inhibitor, pembrolizumab, without any documented IRAE.

Discussion: Hypophysitis is a distinctive side effect of CTLA4-blocking antibodies establishing a new type of autoimmune pituitary disease. The exact pathophysiologic mechanism is not fully delineated but involves T cell activation in the pituitary gland expressing CTLA4 receptors and activation of the classical complement pathway leading to a type II hypersensitivity reaction and pituitary infiltration and gland destruction. It seems to be dose-dependent, occurs more frequently in males, with onset of symptoms 6-12 weeks after initiation of therapy. It can be life threatening if not promptly recognized due to secondary adrenal insufficiency. Patients on CTLA4-blocking antibodies with symptoms suggesting hypophysitis should promptly undergo pituitary MRI and pituitary function testing. If hypophysitis is confirmed, high dose glucocorticoids should be administered as well as replacement hormone therapy for the other axes affected. Hypopituitarism is rarely reversible, and prolonged or lifelong substitutive hormonal treatment is often required. Some studies suggest that development of this IRAE may predict survival of patients with metastatic melanoma.

References

ILLINOIS CLINICAL VIGNETTE POSTER FINALIST - PALLAVI PRADEEP, MD

To B, or not to B

Authors: Pallavi Pradeep, MD, Maryam Saeed, MD, Renato Alcaraz Jr, MD

Introduction: Wernicke’s encephalopathy (WE) is an acute neuropsychiatric derangement caused by Vitamin-B1 (Thiamine) deficiency. It is typically described in alcoholics.

Case Presentation: A 49-year-old Hispanic woman without a significant past medical history was admitted with abdominal pain, intractable nausea, and vomiting, two weeks following a diagnosis of infectious mononucleosis. An upper endoscopy was negative. She was discharged on symptomatic treatment with pantoprazole and sucralfate. Four weeks later, she presented with worsening gastrointestinal symptoms and a new complaint of blurry vision. Physical exam revealed decreased visual acuity with dysconjugate gaze. Brain CT and MRI were unremarkable. Outpatient ophthalmology consultation was advised. Four days later, she reported to the ophthalmologist with worsening vision, headaches, distal tingling/numbness, gait instability, mildly impaired cognition, in addition to the lingering gastrointestinal symptoms. The ocular examination revealed visual acuity restricted to counting fingers, reactive pupils, large central scotoma, restricted abduction/adduction, and bilateral horizontal and vertical nystagmus. She was readmitted with a suspicion of Miller-Fischer variant of Guillain-Barre.

She denied alcohol consumption, tobacco or recreational drug use. She reported 40lb weight loss in 5 weeks. On physical exam, strength was normal, with decreased distal vibration sensation, absent ankle reflexes, and ataxic gait. Anti-GQ1b antibodies and CSF studies were unremarkable. Further testing revealed negative/normal HIV antigen/antibody, vitamin B12 level, glycohemoglobin A1c, Lyme antibodies, angiotensin converting enzyme, ANA and serum protein electrophoresis. Serial brain MRI showed FLAIR hyperintensity in both thalami, dorsal medial brainstem along the mammillary bodies, and the third periventricular white matter. Thiamine deficiency was strongly suspected. A vitamin-B1 level was sent out and she was empirically started on large dose IV thiamine. The thiamine level returned very low at 24nmol/l (70-180nmol/l) confirming WE. Her symptoms began to resolve within 12 hours of thiamine infusion.

Given no preceding history of alcoholism, a workup for other causes of thiamine deficiency was pursued. Celiac serology was negative and stool pancreatic elastase was normal. CT chest/abdomen/pelvis and colonoscopy did not reveal any malignancy or other obvious pathology. Thiamine deficiency was attributed to her prolonged history of vomiting, decreased intake, in combination with the recently prescribed pantoprazole and sucralfate which contributed to malabsorption.

Discussion: WE is defined by the clinical triad of confusion, ophthalmoplegia, and ataxia typically related to alcoholism. From 1867-2014, 623 cases of WE were described in non-alcoholics. Among these non-alcohol related cases, the classic triad was observed in only a third of the patients. We should have a high index of suspicion for thiamine deficiency in situations of unbalanced nutrition that has lasted for 2 to 3 weeks to avoid misdiagnosis of the life-threatening WE or irreversible brain damage (Korsakoff syndrome). This should prompt immediate administration of large doses of thiamine parenterally even prior to laboratory confirmation.

References

A Rare Cause of Recurrent Aspiration Pneumonia

Authors: Faisal Abdulameer, MD. Paul Vishesh, MD

Introduction: Esophageal microperforation is rare and difficult to diagnose and treat, and the causes have been rarely described in literature. Lymphadenopathy of the mediastinum causing bronchoesophageal (BEF) or tracheoesophageal (TEF) fistula is one of the potential causes of esophageal microperforation. The leading causes for TEF and BEF are malignancy and trauma. This case is an uncommon and unusual presentation of an acquired BEF caused by a broncholith that predisposed to esophageal microperforation.

Case Presentation: A 52 year old female with no significant past medical history had recurrent right lower lung abscesses for three times in two years manifesting as productive cough with fevers and night sweats that failed to respond to oral antibiotics. On initial evaluation, CT chest was evident for calcific mediastinal, hilar, and subcarinal lymphadenopathy as well as a right lower lung lobe abscess. Further evaluation with bronchoscopy showed narrowing of the superior segment of the right lower lung bronchus due to a broncholith causing partial compression. Fungal and TB serologies from bronchoalveolar lavage were persistently negative and the patient was responding only to intravenous piperacillin-tazobactam every time. After the third recurrence, a barium swallow showed a broncho-esophageal fistula communicating the mid esophagus to the right lower lobe bronchus explaining the recurrence of abscesses and the association with food intake.

Discussion: A broncholith is a calcified material within the lumen of a bronchus formed when a calcified peribronchial lymph node erodes the bronchus. Broncholiths are usually associated with long standing foci of necrotizing granulomatous lymphadenitis. The vast majority of broncholiths are composed of calcium phosphate (85-90%) or calcium carbonate (10-15%).

Although rare, the literature described broncholiths originating from Nocardia or Histoplasmosis infections but they also originate within intrathoracic tissues previously infected with TB. The typical clinical manifestation is chronic cough, fever, hemoptysis, chest pain, and wheezing. Occasionally, a broncholith can be expectorated while coughing. Recurrent pneumonias may also develop if there is a distal obstruction, like in our case.

One of the rare complications of broncholiths is BEF which is a congenital or acquired communication between the bronchus and the esophagus. The vast majority of acquired BEF cases are caused by malignancy or trauma, and rarely infections. In our case, it was secondary to an esophageal microperforation caused by calcified subcarinal lymphadenopathy, the etiology of which could not be identified.

The suggestive CT findings are partial or complete bronchial obstruction associated with either an endobronchial or peribronchial calcified nodule. The major challenge is to identify endobronchial nodules as thicker CT sections result in a volume averaging artifact of broncholiths. For that reason, higher resolution CT scans have better capability of identifying broncholiths. Treatment options include either continuous observation, removal via bronchoscopy or surgery in more complicated cases.

References

Sneezing leading to Subcutaneous Emphysema and Pneummediastinum in a Navy recruit

Authors: Mohammed Al Tameemi, Muhammad Khan, Ashley Wang, Internal medicine, Chicago medical school at Rosalind Franklin University of Medicine and Science, North Chicago, Illinois.

Introduction: Spontaneous Subcutaneous Cervical Emphysema (Air in subcutaneous tissue) and Pneumomediastinum (Air in mediastinum) due to rupture of the mucosa of the piriform sinus is an extremely rare and dangerous complication of sneezing while obstructing both nostrils.

Case Presentation: 19 years old male Navy recruit with no known co morbidities, presented urgently with sudden onset of neck pain that started after he sneezed with his nose and mouth partially closed. Subsequently, he developed neck swelling, dysphagia and dysphonia. He denies chest pain or dyspnea. There was no history of underlying lung disease, allergies, trauma, weight lifting, smoking or drug intake. His family history is significant for a younger sibling with “air in the orbit” after sneezing with a closed mouth that resolved spontaneously.

On physical examination, patient was alert and not in any respiratory distress. He has a normal body habitus. There was subcutaneous swelling and palpable crepitus on anterior aspect of neck and chest.

Lab results including CBC and CMP were unremarkable. CT neck and thorax showed diffuse neck subcutaneous emphysema and diffuse pneumomediastinum but no pneumothorax or pulmonary emphysema. Flexible fiber optic laryngoscope shows an erythematous, mildly edematous, left hypopharyngeal wall.

Patient was hospitalized and received prophylactic antibiotics to decrease the risk of superinfection and progression to cervical cellulitis. Patient improved symptomatically over the course of stay with resolution of pain, dysphonia and cervical emphysema. A follow up CT scan showed regression of cervical emphysema and Pneumomediastinum. He was discharged on Day 5 with oral antibiotics with limited duty for 10 days. He was also counselled about dangers of sneezing while obstructing both nostrils.

Discussion: Cervical Subcutaneous Emphysema and Pneumomediastinum are often secondary to trauma, infection or surgery with small subsets being primary. Clinical situations predisposing Primary pneumomediastinum are Valsalva maneuvers including sneezing while simultaneously obstructing both nostrils. Rupture of the mucosa of the piriform sinus is extremely rare complication of this type of maneuver leading to air escape in subcutaneous tissue and mediastinum. Complications that can follow includes Mediastinal and Cervical superinfection, Pneumothorax and Tamponade. This case illustrates the fact sneezing while simultaneously obstructing both nostrils is a dangerous maneuver and should be avoided as it can lead to numerous complications.
**Pylephlebitis Caused by a Liver Abscess**

Authors: Daniel Alcantar MD, Fanny Giron Galeano MD, Christine Junia MD

Introduction: Pylephlebitis is a rare complication associated with an intraabdominal septic process, specifically from a region drained by the portal venous system. It is defined as thrombophlebitis of the portal vein and is often reported in association with appendicitis and diverticulitis [1]. It is believed to occur as a result of the spread of the infection into the small vessels that into the portal system [5]. In our case, we describe a case of pylephlebitis that was caused by a left-sided liver abscess.

Case Presentation: A 64-year-old female presented with fevers, chills, myalgia, and loss of appetite. Upon arrival, the patient was found to have a low-grade fever of 100.3 F, a pulse of 103 BPM, and blood pressure of 119/70 mmHg. The physical exam was within normal limits. Hematology and chemistry panel were performed that revealed a leukocytosis of 11.8 K/UL and an alkaline phosphatase elevation of 339 IU/L.

The patient was admitted and continued to be febrile with Tmax of 102.5 F. A CT chest/abdomen/pelvis was performed and revealed a low-density lesion within the left lobe of the liver suspicious for a hepatic abscess and a suspected left segmental pylephlebitis. Intravenous Metronidazole was added to treat for liver abscess. The patient underwent CT-guided aspiration for which 100mL of fluid was aspirated and was sent for cytology and culture. Magnetic Resonance Cholangiopancreatography (MRCP) was performed and a left portal vein thrombus re-confirmed pylephlebitis.

The patient was inevitably discharged home with long-term antibiotics as well as anticoagulation.

Discussion: The diagnosis of pylephlebitis is challenging as there is a broad differential diagnosis to consider. Patients with pylephlebitis may present with non-specific clinical manifestations such as abdominal pain, nausea, vomiting, and fever [4]. Imaging via CT scan (95% sensitivity) and/or ultrasound (51% sensitivity) are utilized to confirm the diagnosis [1].

When considering pylephlebitis, empiric antibiotic coverage for polymicrobial infection, targeting gram-negative aerobes and anaerobes should be initiated. Antibiotics such as metronidazole and a third-generation cephalosporin or fluoroquinolones can be used[9]. Antibiotic therapy is modified according to blood culture results. Treatment can be extended for 4 to 6 weeks. Anticoagulation is also given to prevent thrombus extension. Multiple studies have demonstrated superior outcomes and decreased mortality in patients that were treated with timely anticoagulation and antibiotics versus patients that were treated with antibiotics alone [1, 3, 6].

To our knowledge, there are only 2 cases of liver abscesses as an etiology for pylephlebitis [1]. We believe the presence of the liver abscesses may have extended into a sub-segment of the left portal vein. This case highlights the incidental finding of pylephlebitis as a complication of vast gastrointestinal pathologies. Physicians should be aware of its possibility since it alters the treatment and potential complications for the patient.

References

A Curious Case of Keytruda-induced Crohn’s Disease

Authors: Mirza Ali MD1, Johnathan Fagg1, Priyanka Parajuli1, Mukul Bhattarai1, Southern Illinois University School of Medicine, 1 Department of Internal Medicine

Introduction: Keytruda or pembrolizumab is a humanized monoclonal antibody targeting PD-1 (programmed cell death protein 1) receptors. PD-1 is an immune checkpoint protein which regulates T-cell apoptosis and guards against the development of autoimmunity. PD-1 inhibition, therefore, leads to immune activation against cancer cells.1 It was initially approved by the FDA in 2014 for the treatment of advanced melanoma.2 Since then it has been approved for treatment of non-small cell lung cancer, renal cell cancer, cervical cancer, squamous cell cancers of the head and neck, and Hodgkin’s lymphoma.1 However despite its clinical efficacy, it has been found to be associated with a wide variety of serious autoimmune reactions.

Case Presentation: A 65-year-old male with a past medical history of prostate cancer status post resection, metastatic melanoma presented with approximately two week history of diarrhea. He reported having 8-10 non-bloody, non-mucoid bowel movements per day. He denied fevers, nausea, or vomiting but reported having lost approximately 13 pounds since onset of diarrhea. He denied any recent travel nor did he consume any new foods. Of note, he had been on pembrolizumab for approximately 4 months by the time he presented.

Vitals upon admission were stable and initial labs were consistent with electrolyte abnormalities reflective of diarrhea. CT abdomen without contrast showed dilated loops of bowel with mesenteric lymphadenopathy. Stool analysis was positive for WBCs and occult blood. Stool osmolar gap was consistent with secretory diarrhea. *Clostridium difficile* toxin by PCR was negative. Stool culture was negative for *Salmonella, Shigella, Campylobacter*, and *Escherichia coli* 0157:H7. Hypovolemia and electrolyte abnormalities were corrected. Ciprofloxacin and metronidazole were started for empiric coverage for bacterial enteritis. EGD and colonoscopy were performed and findings were initially concerning for Crohn’s disease. Notably, there was severe chronic inflammation of the duodenum, terminal ileum, and ileocecal valve, in association with crypt abscesses, cryptitis, lymphoplasmacytosis, and distortion of villous architecture. He was started on prednisone and his symptoms improved over the course of one week. Ultimately, he was discharged on an extended prednisone taper over 40 days. EGD and colonoscopy were repeated as outpatient approximately two months after discharge and showed resolution of the aforementioned findings.

Discussion: Here we described an unusual case of pembrolizumab-induced Crohn’s disease in a patient with metastatic melanoma. Autoimmune reactions due to immune checkpoint inhibitors are uncommon but can be life-threatening in some cases if they go unrecognized. In medical literature, pembrolizumab has been found to be associated with multiple autoimmune reactions such as uveitis, pneumonitis, myocarditis, gastritis, acute liver failure, and inflammatory arthritis.3-8 Clinicians should remain vigilant for complications as early recognition and appropriate management is critical and failure to do so may result in delayed care and increased morbidity and mortality.

References


ILLINOIS CLINICAL VIGNETTE POSTER FINALIST - FATIMA AYUB, MBBS

First reported case of Leptospirosis from a ground hog

Authors: Fatima Ayub, MD, Syed Azharuddin, MD, Amir Khan, MD.

Introduction: Leptospirosis is one of the most prevalent zoonotic diseases worldwide, caused by the pathogenic spirochetes, Leptospira. Despite its wide spread distribution, leptospirosis is still underreported. Most of the cases had rodents, rats or dogs as an intermediate carrier but not a single case of leptospirosis in humans caused by groundhog has been reported thus far.

Case Presentation: A 47-year-old Caucasian male presented to the emergency department with complaints of fever, abdominal pain, diarrhea and skin rash for two days.

He went on a camping trip in a local Illinois park, after which he developed fever in the range of 101F-103F, associated with rigors and chills. He also noticed the appearance of a localized, lower abdominal petechial rash with the onset of fever. He also complained of generalized, crampy abdominal pain, associated with nausea. He denied any recent travel outside the United States, rat exposure, flea or tick bites but on further questioning he reported skinning a groundhog two weeks ago.

On arrival, he was hypotensive (90/60mmHg), tachycardiac (120 beats/minute) and febrile (102F). On physical exam, he was ill appearing with scleral icterus, mildly tender abdomen with a blanching lower abdominal petechial rash. His initial abnormal labs include platelet count of 101,000 cells/microliter, creatinine of 2.8 mg/dL and a total bilirubin of 4.5 mg/dL. AST, ALT and alkaline phosphatase were all increased to the levels of 48, 119 and 278 U/L, respectively. During the second day of hospitalization, he was intubated due to development of the acute respiratory distress syndrome. His initial blood cultures were unremarkable. Although his initial Leptospira IgM was negative, he was empirically started on IV doxycycline based on his history of exposure to groundhog, and the heightened suspicion for leptospirosis. Four days later, Karius Digital culture revealed 10,000 copies of Leptospira Kirschneri. He clinically improved within 48 hours of initiation of doxycycline and was discharged on a 7 days course of oral doxycycline.

His repeat labs showed clearance of the organism on Karius digital culture with a positive Leptospira serology. He had complete resolution of his symptoms on the follow up clinic visit.

Discussion: On review of literature, the above described case is the first reported case of Leptospirosis with the only risk factor being a groundhog exposure. Majority of patients with leptospirosis present with non-specific symptoms like fever, myalgia, headache or abdominal pain. Weil’s disease is the severe form of leptospirosis which usually manifests as a classic triad of jaundice, renal failure and pulmonary or brain hemorrhage. A high clinical suspicion is required for the diagnosis of Leptospirosis. An early negative Leptospira IgM should not be considered absolute and should always be rechecked after 7-10 days. Docycycline or penicillin usually provides the best coverage against leptospira.
'Air in My Stomach' - A Case of Emphysematous Gastritis

Authors: Parth M. Desai, DO; Afsana Asharaf, MD; Mohamed A. Elkhouly, MD; Xavier Andrade Gonzalez, MD, Nisar Asmi, MD; Christian Trujillo, MD; Maryam Sanati, MD

Introduction: Emphysematous gastritis (EG) is a rare type of phlegmenous gastritis with less than two hundred cases reported in literature. It is described as presence of air in the gastric wall secondary to a bacterial infection and has been associated with a high mortality rate of up to sixty percent. Most of these cases are preceded by an insult causing gastric mucosal disruption. Here, we report a unique case of emphysematous gastritis in an autistic adult with possible source of infection being the ingestion of foreign bodies.

Case Presentation: A 58-year-old gentleman with known past medical history of Autism, peptic ulcer disease and pyloric stenosis was brought in by his mother due to acute onset of diffuse abdominal pain and melena of one day. On presentation, patient was afebrile and hemodynamically stable. Abdominal examination was concerning for a distended, diffusely tender belly with absent bowel sounds. Labs revealed leukocytosis of 13,200 cells/cubic millimeters. A computed tomography (CT) scan showed distended stomach with gas within the wall without pneumoperitoneum or portal venous gas, compatible with emphysematous gastritis (Fig. 1). The patient was placed on nothing per oral orders and started on total parenteral nutrition, intravenous proton pump inhibitors and broad spectrum intravenous antibiotics. Patient obtained repeat CT abdomen 8 days after onset of symptoms, which showed complete resolution of the gas. An esophagogastroduodenoscopy was done which revealed a superficial ulcer (Fig. 2) in the duodenal region and the presence of three foreign bodies in the stomach, that was successfully removed (Fig. 3-4). The patient was then started on a diet and discharged home.

Discussion: EG is a rare bacterial infection that presents with air in the gastric mucosa, usually caused by organisms such as C. perfringens, Escherichia coli, Enterobacter aerogenes, Proteus vulgaris, Pseudomonas aeruginosa, non hemolytic streptococcus and Staphylococcus. One of the prime functions of the gastric mucosa is to act as a barrier against infection. However, previous gastroduodenal instrumentation and corrosive acid ingestion can disrupt this barrier, thus, creating a portal for microorganism entry. We speculate that the source of infection in our patient was the foreign bodies that he ingested. The presence of a duodenal ulcer and slow motility increased the risk of EG by acting as a gateway for the microorganism to enter the gastric mucosa. A computed tomography scan showing cystic pockets of streaks of air within the gastric wall can help clinch the diagnosis. Conservative management is the mainstay of therapy with early initiation of broad spectrum antibiotics demonstrating good clinical outcomes. In the acute setting, the role for surgery has not been well defined. Surgery including gastric resection is often reserved for patients who deteriorate despite optimal medical management or when there is evidence of infarction or perforation.
A rare case of upper extremity deep venous thrombosis due to aortic arch compression

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Introduction: Upper extremity deep venous thrombosis (UEDVT) is much less common than lower extremity DVT1. The well-documented risk factors are central venous catheters, pacemakers, cancer, or other thrombophilic conditions1. UEDVT can also occur from repetitive upper limb activity in an athlete or an individual with thoracic outlet anatomic abnormalities. The absence of these risk factors may suggest an uncommon etiology. Here, we report a rare case of UEDVT due to aortic arch compression.

Case Presentation: A 70-year-old female presented with two days of left neck swelling. Her past medical history was significant for a single-vessel CABG with bioprosthetic aortic valve replacement five months prior. She was on aspirin post-operatively. She had no personal or family history of thrombosis. She does not engage in any repetitive arm activities. Additionally, she has never had a central venous catheter.

She underwent venous Doppler imaging of her upper extremities, which showed an acute deep venous thrombosis in the left internal jugular vein. She subsequently underwent CT chest venogram, which demonstrated an aneurysmal and tortuous ascending aorta causing compression of the left brachiocephalic vein, with acute clot in her brachiocephalic vein extending into the left internal jugular vein to the level of the thyroid cartilage.

She was started on anticoagulation with apixaban with resolution of her swelling and referred to cardiothoracic surgery for surveillance of her aorta.

Discussion: Virchow’s triad of hypercoagulability, venous stasis, and vessel wall injury describes the fundamental mechanisms of venous thrombosis. Our patient’s UEDVT is secondary to venous stasis from extrinsic compression by her enlarged and tortuous aorta. A parallel example of this mechanism in the lower extremities is May-Thurner Syndrome, which is most commonly due to left iliac vein compression against the vertebrae by the right iliac artery. We can also compare this case to venous thoracic outlet syndrome, in which the subclavian vein is compressed between the first rib and clavicle.

The management options for symptomatic proximal UEDVT include anticoagulation alone or thrombolysis with stent placement2. Our patient was treated with anticoagulation and had significant improvement in her symptoms within days. Currently, she remains on apixaban with close surveillance.

There are few case reports of UEDVT due to aortic arch compression. In one case, clinicians were unable to place a pacemaker lead due to occlusion of the left brachiocephalic vein from aortic arch compression3. This patient was asymptomatic due to the presence of venous collaterals. In another case, a patient developed an asymptomatic left internal jugular thrombosis due to a right-sided aortic arch aneurysm4. These rare cases and ours serve to illustrate how UEDVT can be a sign of intrathoracic compression and should prompt further evaluation for compression in the absence of other common risk factors.

References


Not one but two, multidisciplinary team is the clue Synchronous diagnosis of intrahepatic cholangiocarcinoma and diffuse large B cell lymphoma

Authors: Karam Khaddour M.D, Veerpal Singh M.D

Introduction: According to the International Agency for Research on Cancer (AIARC) synchronous multiple primary malignancies is defined as the diagnosis of more than one distinct histological and morphological primary malignancy within a period of less than 6 months. It is estimated that the frequency of multiple primary cancers can reach up to 17% in cancer survivors and it could be challenging to establish a presence of multiple malignancies at the same time given that most of them are clinically silent and may appear as the same disease on imaging. We describe a case where effective communication led to the diagnosis of synchronous intrahepatic cholangiocarcinoma and diffuse large B cell lymphoma in a patient presenting with non-specific symptoms and a single hepatic lesion without lymphadenopathy.

Case Presentation: A 70 year old female with no past medical history presented with complaints of abdominal discomfort and unintentional weight loss. Abdominal ultrasound showed ascites with nodular hepatic lesion. Initial Computed Tomography (CT) revealed a heterogeneous lesion which was biopsied and was consistent histologically with intrahepatic cholangiocarcinoma. There were no signs of metastatic disease on whole PET-CT scanning. The patient had paracentesis due to her abdominal discomfort and large accumulative ascites. During microscopic examination of the peritoneal fluid it was noted by pathology and laboratory team that the malignant cells did not resemble an origin of adenocarcinoma of the primary cholangiocarcinoma which prompted them to call for a meeting with multidisciplinary team members involved in the care of this patient. After the meeting and discussion on available literature it was concluded to proceed with flow cytometry and PCR which showed monoclonal rearrangement of the immunoglobulin heavy chain and kappa light chain. Immunohistochemically staining was positive for CD20 and the findings were consistent with diffuse large B cell lymphoma (DLBCL). Given the diagnosis of two different malignancies within 1 month period the patient underwent extensive work up for proper staging of each malignancy. A thorough review of the literature of treatment strategies was presented in multidisciplinary team meetings. After discussion with the patient she was started on Gemfibrozil, Cisplatin and Rituximab. On 6 month follow up the patient showed remission of her DLBCL and no signs of progression of cholangiocarcinoma. Genetic testing was offered which the patient declined.

Discussion: Studies show that in hospital multidisciplinary teams improve patients’ outcome. In our case, effective communication led to the diagnosis of two different primary cancers that presented as one tumor as the ascites was thought initially to be secondary to cholangiocarcinoma. Due to laboratory observations and their call for reconsidering the diagnosis a more comprehensive approach effectively helped uncover diffuse large B cell lymphoma that was treated along with cholangiocarcinoma to achieve the best outcome for the patient.

References

An Ablated Heart and a Bubbled Brain - a delayed complication.

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Introduction: Catheter ablation procedures have emerged as a major tool to treat refractory arrhythmias. The incidence of complications from ablation procedures is less than 3%. In this case report, we present a rare complication due to an ablation procedure with very high mortality.

Case Presentation: A 68 years old female, smoker with BMI-54.68 kg/m2 presented to the emergency department with multiple falls and episodic confusion for 2 days. She denied weakness, numbness, loss of consciousness, seizures, dizziness or prior gait problems. For the past 4 weeks, she had worsening exertional shortness of breath, mid chest tightness on breathing in and occasional dry cough. She was hospitalized a week ago for sepsis secondary to suspected pneumonia. She underwent cryo-balloon ablation with pulmonary vein isolation procedure for persistent atrial fibrillation- 4 weeks ago.

On physical examination, she was hypotensive, febrile, tachypneic and decreased breath sounds with scattered crackles were noticed at both lung bases; the remainder of the examination were unremarkable. Her significant labs included lactic acidosis, elevated procalcitonin (28.59), but no leukocytosis. Chest X ray showed worsening pulmonary infiltrates with cardiomegaly. Electrocardiogram, echocardiogram and initial CT brain without contrast were unremarkable. Further her hospitalization course was complicated with intermittent confusions, transient left hemiparesis resolved in 24 hours and worsening hypoxemia- requiring 6L/min of nasal oxygen. Infectious workups including QuantiFERON gold, HIV were unremarkable. Contrast enhanced CT chest showed mediastinitis with focal air collection in the posterior mediastinum and mild dilated oesophagus, concerning for esophageal or bronchial fistula/leak. Further MRI brain with contrast revealed acute infarctions in the high-bilateral frontoparietal regions consistent with air embolism. But esophageal fluoroscopy studies didn’t show leakage. She underwent explorative thoracotomy and an Atrio-esophageal fistula (AEF) was identified intraoperatively and repaired. Her post-operative period was uneventful, and her symptoms resolved.

Discussion: AEF after cryo-balloon ablation is rare complication, with incidence of <1 per 10,000 cases. The mean interval from the procedure to the clinical presentation is in range of 3 to 60 days. Presenting symptoms include fever, neurological deficits and haematemesis. In our patient, the presentation was atypical, initially suspected as pneumonia, later identified as mediastinitis.

AEF has a very high mortality rate of 60-70% likely from cerebral air embolism, severe sepsis and massive gastrointestinal bleeding. The important teaching points are:

- delayed presentation of AEF
- To avoid TEE and upper GI endoscopy, because it can lead to widespread air embolism.
- Early surgical repair prevents mortality.

References

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Coxsackie B Infection Presented as Severe Acute Pancreatitis and Pneumonia with Bilateral Pleural Effusion in a Navy Recruit

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Introduction: Coxsackie viruses are enteroviruses belong to the Picornaviridae family. Coxsackie group B viruses have been associated with many clinical syndromes including pleurodynia, pleural effusion, pneumonitis, pericarditis, myocarditis, and pancreatitis. There are very few reports and studies concerning Coxsackie virus infection with pulmonary involvement or pancreatitis.

Case Presentation: A 18-year-old male previously healthy Navy recruit from Arizona, fell ill with sore throat, cough, pleuritic chest pain, and subjective fever. Four days later, he developed severe epigastric radiating to his back associated with nausea and vomiting that prompted medical attention. The patient had no personal or family history of gallstone or liver disease. He denied using any medications, alcohol or recreational drug use. He reported sick contacts.

On physical examination, his temperature was 100.5°F and heart rate 112bpm. He had coarse breath sounds and severe tenderness to palpation in the epigastric region. There was no rash on the body.

Laboratory investigation showed leukocytosis (17,000 K/uL), elevated lipase to 4268 U/L. Renal and liver functions were normal. CT chest showed scattered ground glass opacities in both lung fields. CT abdomen showed pancreatic edema with peri pancreatic fluid collection consistent with acute pancreatitis. Recruit was admitted for pancreatitis and pneumonia.

Etiological workup showed normal ultrasound with no gall stones. No hypertriglyceridemia, or hypercalcemia. Autoimmune pancreatitis was ruled out with negative IgG-4. His CPK and Troponin were normal. MRCP was unrevealing for structural pancreatic abnormality including pancreatic divisum. Aspergillus antigen, Leptospira, and Legionella were negative. Blood and sputum culture were negative. Viral panel were negative for Influenza, Ebstein Barr virus, Cytomegalovirus, Mumps, Varicella zoster and Coxsackie A, however antibody titers against Coxsackie B type 5 were markedly elevated.

The patient was treated symptomatically with IV hydration and analgesics. Two days after admission, his abdominal pain worsened and he developed hypoxemic respiratory failure requiring supplemental oxygen. Repeat CT scan showed worsening of severe acute pancreatitis with bilateral pleural effusions. Supportive treatment was continued. The patient had gradual clinical recovery over nine days with resolution of abdominal pain, cough, chest pain, and return of appetite. Leukocytosis resolved and lipase trended down to normal. Recruit was discharged in stable condition.

Discussion: Acute Pancreatitis has diverse etiologies with viral etiology involving only a small subset. Imrie et al carried out a prospective study on 116 patients with acute pancreatitis with incidence of idiopathic pancreatitis in this study was 5.2% (six patients). Among them, five patients exhibited significant rising antibody titers to coxsackie B or mumps. There are relatively few case reports concerning Coxsackie virus infection with pulmonary involvement and incidence remains unknown. Our case illustrates the importance of considering Coxsackie B infection as a rare cause of acute pancreatitis especially with concomitant respiratory symptoms.
Takotsubo cardiomyopathy with rapid recovery following induction anesthesia

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Introduction: Takotsubo cardiomyopathy is characterized by transient regional systolic dysfunction of the left ventricle, mimicking myocardial infarction, but without obstructive coronary artery disease. We present a 48 year old man who presented for an ear surgery, developed takotsubo cardiomyopathy following induction anesthesia, and had a rapid recovery.

Case Presentation: 48 year old man with cholesteatoma of right ear and hypertension was admitted for tympanomastoidectomy. Pre-operative blood pressure was 113/83 mm Hg and heart rate was 69 per minute. A few minutes into induction anesthesia with midazolam, fentanyl, lidocaine and propofol, patient developed hypotension and transient bradycardia: blood pressure nadir was 70/50 mm Hg and heart rate nadir was 60 per minute. Several rounds of intravenous phenylephrine, epinephrine and ephedrine were given and isotonic fluid was infused leading to partial improvement in blood pressure. Cardiac troponin at the time of event was 0.036 ng/mL, 1.052 after 6 hours and 0.133 after 19 hours. ST segment depressions were observed in the monitor inside operating room, however, electrocardiogram done shortly after showed few premature ventricular complexes but no ST segment-T wave changes. Potassium was 2.9 mEQ/L. Transthoracic echocardiogram showed reduced left ventricular ejection fraction (LVEF 40%) with hypokinesis of the apex, anterior, inferior, septal and lateral walls of the myocardium. A coronary angiogram revealed no coronary artery disease. A diagnosis of takotsubo cardiomyopathy was made and patient was transferred to coronary care unit. Extended release metoprolol was begun once blood pressure was permissive. Transthoracic echocardiogram repeated 3 days later showed low normal LVEF (50-55%), without regional wall motion abnormalities.

Discussion: Pathogenesis of takotsubo cardiomyopathy is thought to be a catecholamine excess causing coronary artery spasm and microvascular dysfunction, leading to myocardial stunning. The usual presentation consists of ST segment elevation in anterior precordial leads, elevated cardiac troponins, and ventriculography revealing LV apical hypokinesis and basal hypercontractility. The temporal pattern in this case reveals bradycardia and hypotension minutes after intravenous midazolam, fentanyl, lidocaine, and propofol. Each of these agents have been independently recognized to cause both hypotension and bradycardia. Following this, patient was appropriately resuscitated with catecholamines and sympathetic agonist. While it can be argued that patient could have had a catecholamine surge in anticipation of or during anesthesia, leading to takotsubo cardiomyopathy, a more plausible explanation could be the catecholamine excess patient was exposed to during the resuscitation process. Review of other cases of takotsubo cardiomyopathy following anesthesia induction revealed that most of them were concurrently exposed to catecholamines too. This case begs awareness among physicians about this iatrogenic entity. Also, should we use alternative agents such as terlipressin for perioperative blood pressure management? Recovery generally occurs in 1 to 4 weeks. The rapid recovery of EF in less than 3 days is something unique in this case.
An uncommon presentation of pyogenic liver disease caused by Streptococcus pneumoniae

Introduction: Pyogenic liver abscesses (PLAs) are uncommon conditions that occur as complications of gastrointestinal infections or bacteremia. Its etiology is normally polymicrobial and the spectrum of microorganisms are related to patient comorbidities and local epidemiology. Few case reports have described Streptococcus pneumoniae as etiology of liver abscesses but none of them were in the United States. We present a case of pneumococcal liver abscess in the absence of respiratory symptoms.

Case Presentation: 67-year-old female with type-2 diabetes, asymptomatic liver cysts, and a recent diagnosis of pancreatic adenocarcinoma with post endoscopic retrograde cholangiopancreatography (ERCP) and stent placement two weeks prior to admission, presented to the hospital with one-week history of right upper quadrant abdominal pain and subjective fever. On physical examination, she was found febrile, tachycardic and hypotensive. Laboratory workup showed leukocytosis and elevated total bilirubin (direct predominance), alkaline phosphatase, aspartate transaminase, gamma glutamyl transferase and lactate. An abdominal computed tomography (CT) scan described an increase in the size of one of the hepatic cysts in the posterior segment of the right lobe surrounded by edema (Image 1), highly concerning for abscess formation. The patient was admitted with the diagnosis of sepsis and intravenous (IV) fluids and broad-spectrum antibiotics with vancomycin, ceftriaxone and metronidazole were rapidly started. Subsequently, liver abscess was drained and culture was positive for Streptococcus pneumoniae. After 48 hours from admission, fever and abdominal pain resolved, vital signs normalized and antibiotics were switched to levofloxacin and ampicillin-sulbactam. An abdominal CT scan control showed a decrease in size of the fluid collection (Image 2) and blood cultures remained negative after five days from admission. Given good clinical improvement, antibiotics were switched to amoxicillin-clavulanic acid and the patient was discharged to complete a six-week of antibiotics therapy. Patient was seen in the post-hospital follow up four weeks after discharge and was found asymptomatic.

Discussion: PLAs are a rare but important cause of morbidity and mortality around the world. Its incidence in the United States is 3.6 cases per 100 000 population and has been slowly growing over the last decade. The microbiological spectrum is broad and varies mainly depending on the local epidemiology and host risk factors. Streptococcus pneumoniae has been rarely associated with intra-abdominal infections including hepatic and biliary tree infections and recently case reports have described an association with PLAs. The formation of PLAs is usually explained by spreading from the biliary tree, portal vein, or hepatic artery, but also by direct extension from another abscess, trauma or through biliary procedures as ERCP. Treatment includes drainage along with IV antibiotics with a complete resolution in most of the cases within few weeks to months. It is believed that the initial infectious source might be in the respiratory tract with a subsequent bacteremia in susceptible patients for invasive pneumococcal infection as diabetics. Bacteremia will posteriorly seed in the liver or biliary tract leading to abscess formation.

References
"Isabel" To The Rescue!

Authors: J. Zabala-Genovez, M.D.; Edwarda Golden, M.D.; Farah Ciftci, M.D. Transitional and Internal Medicine Service, Loyola Medicine MacNeal Hospital.

Introduction: The ability to accurately estimate probability to make a diagnosis is one of the most important qualities of a good physician. Differential diagnosis generators may be helpful in creating more accurate differential diagnosis in complex cases based on true probability.

We are presenting a case where the differential diagnosis generator guided the team to recognize the illness script.

Case Presentation: A female of 70-years with a history of osteoarthritis presented with four weeks of fever, weight loss, night sweats, and fatigue. She was hypotensive, febrile, tachycardic and tachypneic. She was initially managed per sepsis protocol. Laboratory data revealed WBC 2.7 K/UL, Hgb 9.8 g/dL, PLT 77 K/DL, AST 71 IU/L ALT 49 IU/L, CRP 12 mg/dL, ESR 41 mm/hr. Ferritin 7138 ng/mL, LDH 570 IU/L. She continued to be febrile, pancytopenic and generally ill for the first 48 hours despite IV antibiotics. A contrast-enhanced CT of the abdomen revealed splenic hypodensities; laboratory workup for Q-fever, RMSF, salmonella, and Lyme’s disease was sent to evaluate for secondary hemophagocytic lymphohistiocytosis (HLH).

Lack of clinical and laboratory improvement prompted us to utilize Isabel Healthcare Differential Diagnosis (DDX) Tool, a resource available in our institution. Our keywords generated ten differential diagnosis including Brucellosis. Our patient later admitted consuming homemade cheese on her last visit to Mexico. Brucella serology, blood cultures, and a liver biopsy were obtained. Fever resolved 48 hours after doxycycline was initiated. Blood cultures grew Brucella spp. Currently, our patient remains symptom-free and all laboratory abnormalities have normalized.

Discussion: Medical decision making relies on pattern recognition of illness scripts accumulated over time. Illness scripts can be contaminated by our own heuristics and biases. Complex and rare conditions like HLH secondary to Brucellosis may require more analytical decision making with the establishment of true probability. Historically, we referred to textbooks for the DDX for each symptom. Nowadays, we have more sophisticated DDX generators that include epidemiological data, clinical and laboratory findings to generate a DDX list. In a study, the pooled accurate diagnosis retrieval rate for DDX tools was as high as 0.70. To be able to utilize a DDX generator properly, it is crucial to identify significant and clinically relevant data. The expert physician goes through a series of hypothesis refinement during the process of solving a complex case and the search might be more refined to find the most probable etiologies. More studies are needed to test these systems’ ability to provide a final diagnosis. At this time, DDX generators may be useful resources for clinicians and it may also increase the medical knowledge.
Trust Your Gut: A Case of Fever of Unknown Origin

Authors: Daisy Zhu, MD

Introduction: Lemierre’s syndrome classically presents as thrombophlebitis of the internal jugular vein after oropharyngeal infection. A rare abdominal variant of Lemierre’s syndrome has been implicated as a cause of pyelitis, which is defined as thrombophlebitis of the portal venous system. Patients with the abdominal variant of Lemierre’s syndrome may develop complications such as hepatic abscesses and bowel ischemia.

Case Presentation: A 73-year-old male with a past medical history of melanoma resected with clear margins, hypertension and hyperlipidemia presented to the emergency room with a seven-week history of fever. His symptoms started two days after returning from a trip to Arizona. He initially developed a non-productive cough, followed by high-grade fevers, chills and drenching night sweats. He was physically active at baseline, but endorsed profound fatigue and generalized body aches since symptom onset. He denied associated nausea, vomiting or diarrhea. Prior to presentation, he was evaluated by an Infectious Diseases specialist. Workup was notable for increased mesenteric adenopathy on CT chest/abdomen/pelvis and negative Coccidioides antibody titer. Despite empiric therapy with Fluconazole for presumed coccidioidomycosis, the patient’s symptoms worsened. He presented to emergency room with fever of 101.5 °F (38.6 °C), white blood cell count of 19,100 cells/μL with 85.1% PMNs, lactate 2.3 mmol/L, ESR 92 mm/hr and CRP >300 mg/L. He denied hiking, rafting or having any contact with animals while in Arizona. He had not traveled outside the USA in ten years and denied sick contacts. Repeat imaging with CT and MRI showed extensive portal vein thrombosis extending into the hepatic and superior mesenteric veins, moderate abdominal ascites and hepatic abscesses. The imaging findings raised concern for an abdominal variant of Lemierre’s syndrome. Paracentesis was performed and the ascitic fluid grew 4+ WBCs with no organisms identified. The patient was started on therapeutic anticoagulation and Piperacillin-Tazobactam given suspicion for an anaerobic source. Six days later, blood cultures obtained on admission returned positive for Fusobacterium nucleatum. Notably, the Brucella IgM titer returned positive; however, Infectious Diseases specialists believed the result was falsely positive as blood cultures did not identify the organism. The patient’s fevers resolved and he was discharged with Ertapenem 1 g daily to complete six weeks of therapy at an outpatient infusion center.

Discussion: Fusobacterium spp. is an anaerobic, gram-negative rod found in the gastrointestinal tract. Its thrombogenic nature has been documented in literature as a common pathogen for Lemierre’s syndrome. However, the abdominal variant of Lemierre’s syndrome is extremely rare. To date, there are fewer than ten published case reports, 4 of which had preceding oropharyngeal infections. This case highlights the importance of considering intra-abdominal pathogens such as Fusobacterium spp. in the setting of bacteremia and portal vein thrombosis, as early diagnosis and treatment have been shown to result in fewer complications and improved outcomes.

References

Multaq and the Mysterious Effusions: A Case of Drug-Induced Lupus Secondary to Dronedarone

Authors: Ishan Gohil MD, Lyndsey Booker MD, Laura Hampton MD, Cassidy Menard MD

Introduction: Drug-Induced Lupus (DIL) is a rare systemic autoimmune condition due to autoantibody generation secondary to a provoking medication. Some features of DIL overlap with systemic lupus erythematosus (SLE), but DIL tends to have more abrupt onset, older age at onset, and spare the kidneys and central nervous system. We present a fascinating case of a patient with recurrent pleural and pericardial effusions secondary to drug-induced lupus from dronedarone.

Case Presentation: A 70-year-old female with distant history of chest radiation and atrial fibrillation presented to our hospital due to worsening dyspnea, effusions, weight loss, and tamponade physiology on echocardiogram. Ejection fraction was 45-50% with focal hypokinesis of the inferior and anterolateral walls. Over the past nine months, she had recurrent pleural effusions requiring multiple thoracenteses. Outpatient fluid studies demonstrated transudative effusions negative for infection, tuberculosis, or malignancy. Serum ANA titer was 1:160 on one occasion but subsequently negative on repeat testing.

The patient underwent a pericardial window, talc pleurodesis, and pleural biopsy. Fluid studies from both the pleura and pericardium confirmed transudative effusion. Postoperative echocardiogram revealed improved ejection fraction of 50-60%, normal wall motion, and resolution of tamponade physiology. Studies from both serum and fluid including ANA, complement, ANCA, rheumatoid factor, bacterial and fungal cultures, and cytology were all within normal limits. Anti-histone antibody was strongly positive in both serum and pericardial fluid.

Review of patient’s history revealed start of dronedarone therapy approximately 12 months prior to admission, three months before the onset of effusions. A thorough medication review demonstrated no other culprit medication. Dronedarone was stopped and rate-control started with a beta-blocker. Due to persistent output from her pericardial drain and small recurrent pleural effusion, low dose prednisone was started. She had a significant decrease in drain output within 24 hours. The drain was removed, and she was subsequently discharged with outpatient Rheumatology and Cardiology follow-up. Repeat chest radiograph and echocardiogram one month later revealed complete absence of effusions.

Discussion: Drug-induced lupus is rare. Diagnosis is made by a thorough history and physical exam, elucidation of a provoking medication, positive serologic markers, clinical symptoms consistent with lupus, and improvement after drug cessation. Anti-histone antibodies are most strongly tied to DIL and present up to 95 percent of the time with traditionally implicated drugs. Amiodarone has been reported to cause DIL, but from our literature search, this would be the first case of dronedarone-induced lupus. Typical manifestations include arthritis, serositis and cutaneous findings. Hematologic, renal, and CNS abnormalities of SLE are very rare with DIL, consistent with our patient’s presentation. Treatment entails stopping the offending medication. Serious complications can benefit from systemic corticosteroids, which usually provides rapid initial improvement. Prognosis is typically favorable but may take weeks to months until full recovery.

References

The Sheep in Wolf’s Clothing: ST-Segment Elevation, Not Always a Myocardial Infarction.

Authors: Shiv Bagga, MD, Nicole Sbircea, DO, Jennifer Mundell, MD.

Introduction: Rapid recognition of ST-segment elevation (STE) myocardial infarction (STEMI) is imperative; as prompt reperfusion can dramatically reduce the associated mortality and morbidity. However, electrocardiogram (ECG) remains an imperfect diagnostic tool as 60-80% of patients presenting with STE are found not to be associated with STEMI. Indeed, STE may also be seen in patients with early repolarization, acute myo/pericarditis, coronary vasospasm, hyperkalemia, Takotsubo cardiomyopathy, large pulmonary embolism and Brugada syndrome. We present a case of STE due to hypercalcemia, one the more rarely described ECG manifestations of severe hypercalcemia.

Case Presentation: A 60-year-old gentleman with history of squamous cell carcinoma of the tongue, post resection, radiation and chemotherapy with cisplatin was admitted to the emergency department with acute encephalopathy. Laboratory results were pertinent for a blood urea of 74, serum creatinine of 7.4 mg/dl and corrected serum calcium of 15.8. An ECG demonstrated a new STE in V1-V3 with biphasic T waves in V2-V6. PR interval was 216 ms, QTc was 378 ms while QaTc was 200 ms. He denied chest pain, but with concern for an ACS, troponins levels were obtained which were 0.04 and 0.03 ng/ml. Transthoracic echocardiogram showed an ejection fraction of 60% with no segmental wall motion abnormalities. Further workup showed a parathyroid hormone at low end of normal range while parathyroid hormone-related peptide was significantly elevated. Imaging confirmed recurrence of cancer and the patient was referred to oncology for additional treatment. Repeat ECG demonstrated resolution of STE with normalization of serum calcium.

Discussion: The most commonly recognized ECG manifestations of hypercalcemia are QT interval shortening with less common findings being PR prolongation and a diffuse increase in amplitude of the QRS complex. STE is one of the more rarely described ECG manifestation with descriptions limited to case reports and only one case series of patients with moderate to severe hypercalcemia. STE due to hypercalcemia is most commonly seen in the anterior precordial leads and these usually have a scooped appearance with no distinct T waves. The mechanism of STE is proposed to be due to hypercalcemia induced QT shortening essentially pulling the T wave closer to the QRS complex thereby causing a high take-off of the ST segment giving pseudo appearance of a ST elevation. Though QT interval shortening seems counterintuitive in the setting of hypercalcemia, it is related to elevated calcium causing calcium dependent inactivation of the L-type calcium channel activity during phase 2 of the action potential. It is crucial that clinicians consider other causes of STE and be aware that hypercalcemia may be associated with a pseudo-infarct pattern on the ECG. Failure to recognize these mimickers of ACS can lead to inappropriate use of resources and delay appropriate treatments.

References

Biopsy Proven Resolution in Lymphocytic Colitis after Withdrawal of Paroxetine.

Authors: Adil Ghafoor, MD, PGY2, Department of Medicine, St. Vincent Health, Indianapolis, IN., Mazen Alsatie, MD, Department of Gastroenterology, St. Vincent Health, Indianapolis, IN.

Introduction: Microscopic colitis (MC), with its subtypes; lymphocytic (LC) or collagenous (CC) colitis, is diagnosed by random colon biopsy. Most cases are idiopathic but association with medication such as selective serotonin reuptake inhibitors has been reported, primarily through retrospective case-control studies or case reports. Publications have consistently emphasized sertraline’s high likelihood and paroxetine’s intermediate likelihood of this association with MC. For any medication implicated in drug-induced MC, a cause-effect relationship has not been established as the mechanism is not well understood. Nonetheless, few reports indicate biopsy-proven resolution or improvement in the histology after discontinuing the culprit medication. By demonstrating clinical and histological improvement or resolution of MC after discontinuing the implicated medication, we can build a stronger case for the medication as a culprit of MC. Below, we chronicle the diagnosis of diarrhea and drug-induced lymphocytic colitis secondary to paroxetine, with close follow-up confirming clinical resolution and significant histologic improvement upon the discontinuation of paroxetine.

Case Presentation: The patient is a 35-year-old female with a history significant for generalized anxiety disorder, lumbar radiculopathy, and psoriasis who presented to her primary care physician’s office with a 9-month history of diarrhea and a recent 2-week history of intermittent hematochezia. She denied weight loss, fevers, nausea, vomiting, and jaundice. Her medications included meloxicam 15 mg daily as needed for radiculopathy and daily paroxetine 12.5 mg daily for generalized anxiety. Gastroenterological evaluation included serology for celiac disease, stool testing, and unremarkable routine blood work. Colonoscopy revealed one flat 12 mm polyp in the splenic flexure and a second flat 10 mm polyp in the ascending colon, both diagnosed as sessile serrated polyps on histology. Random colon biopsy revealed prominent intraepithelial lymphocytosis consistent with lymphocytic colitis. The colitis was presumed to be due to paroxetine and the patient discontinued this medication. The patient’s diarrhea resolved within 2 weeks of discontinuing the paroxetine. On a follow-up colonoscopy performed 6 months afterwards, a repeat mucosal biopsy revealed mild intraepithelial lymphocytosis, a significant histologic improvement of the lymphocytic colitis.

Discussion: Due to the lack of evidence suggesting a clear mechanism through which a medication can cause MC, cause-effect relationship has not been established. Therefore, implicated medications are categorized as high likelihood, intermediate likelihood, and low likelihood of causing MC. Criteria for this classification process include timing of drug initiation and onset of symptoms, improvement in symptoms with drug cessation, and recurrence of symptoms upon repeated exposure to the drug. Most medications associated with MC are known to be associated with chronic diarrhea. Therefore, demonstrating the resolution or improvement of histologic changes on colonic biopsy after withdrawal of a suspected medication can provide further objective data and build a stronger association with medications and MC as demonstrated in our clinical case with paroxetine.
I’m Weak, Can’t Breathe, Can’t See, and Have RBC’s In My Pee: What Am I?

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Introduction: Sudden, painless, unilateral vision loss can present variably, and the differential diagnosis typically includes ischemia such as retinal artery and vein occlusion, optic neuritis and inflammatory conditions such as giant cell arteritis. We present a unique case of acute, painless, unilateral vision loss associated with renal failure and elevated inflammatory markers suspicious for a systemic vasculitis.

Case Presentation: 64-year-old lady with no significant past medical history presents to the ED with her second episode of sudden, painless, left eye blindness within the week. Additionally, she reported having dry cough, fatigue and anorexia for approximately 2 months, and was diagnosed with a sinus infection on CT scan during that time. She was treated with antibiotics and steroids, with only minor improvement in her symptoms. About 1 week prior to her ED presentation, she woke from sleep and could not see out of her left eye. She ultimately went back to sleep and the vision loss resolved by morning. She was referred to an ophthalmologist who thought it was secondary to retinal artery occlusion. After the second occurrence on the day of presentation, she contacted her ophthalmologist and was instructed to go to the ED. On admission, she met SIRS criteria with leukocytosis and tachycardia, had acute renal failure with a creatinine of 3.64, and an ESR of 107 with a CRP of 13.7. CXR revealed an opacity of the right mid-lung. CT brain revealed mucosal thickening of the ethmoid and sphenoid sinuses bilaterally. Urinalysis revealed trace protein with large occult blood and greater than 100 RBCs. Her renal ultrasound was negative and her FeNa was 4.4. Her Proteinase 3 antibody returned highly elevated concerning for Granulomatosis with polyangiitis. Empiric IV steroids were started, and her vision subsequently returned to normal. Due to an underwhelming response to steroid therapy alone, her steroid dose was increased along with initiating Rituxan and Atovaquon. A renal biopsy was scheduled for confirmation of the diagnosis.

Discussion: Granulomatosis with polyangiitis (GPA), formerly known as Wegeners Granulomatosis, is the most common small vessel vasculitis. It falls into the class of C-ANCA auto-antibodies, also known as anti-PR3 autoantibodies of neutrophil granulocytes. This results in activation of neutrophils and release of metalloproteinase and toxic free radicals, causing destruction and inflammation of small vessels, affecting several organs. GPA typically involves the respiratory and renal systems, as described in this patient; however, less than 20% of cases present initially with ocular symptoms. It is critical then that when a patient presents with unilateral painless vision loss, systemic vasculitides are included in the differential diagnosis as time to initiation of treatment is crucial to preserve vision.
On the fifteenth year of use, my statin gave to me: Forty thousand CPK, 8 days of hospitalization and a pair of unstable legs

Introduction: With a growing population suffering from hypertension and hyperlipidemia, the number of patients who are being co-prescribed antihypertensives and lipid-lowering agents is increasing. Here, we present a unique case of severe rhabdomyolysis in a patient on simvastatin for 15 years without any side effects who developed debilitating proximal muscle weakness after initiation of verapamil.

Case Presentation: A 70-year-old female with history of hypothyroidism, hypertension, hyperlipidemia, and CKD stage IV presented to the hospital with 10 days of progressively worsening proximal lower extremity weakness so severe she was unable to ambulate. An outpatient MRI revealed multifocal areas of moderately intense muscular edema within the pelvis and proximal thighs, suggesting multifocal inflammatory myopathy such as idiopathic polymyositis. She denied any vaccinations or previous musculoskeletal diseases. Her physical examination was unremarkable except for symmetrical proximal muscle weakness of the lower extremities. Her labs included: CPK greater than 42670, creatinine 2.25, TSH 6.43, LDH 2622, ESR 42, CRP 1.1, positive ANA of 1:320, and negative auto-antibodies. Given the severity of her symptoms and concern for polymyositis, high dose steroids were administered. Further history revealed that she began verapamil 120mg daily for hypertension control the previous month. She had been on simvastatin 80mg for the previous 15 years without issues. A muscle biopsy revealed severe myofiber necrosis, suggestive of medication-induced toxicity. Given temporal relationship between initiation of Verapamil and her symptom onset, negative inflammatory and endocrine workup, and confirmed muscle biopsy, we determined that rhabdomyolysis was due to simvastatin toxicity. Simvastatin and verapamil share CYP3A4 pathway where simvastatin concentration increases as verapamil inhibits CYP3A4-based metabolism of simvastatin.

Discussion: Statin-induced rhabdomyolysis, although rare at 0.1%, is a debilitating side effect1. In 1998, a handful of rhabdomyolysis-related deaths prompted the withdrawal of mibefradil and cerivastatin2. CCBs act as CYP3A4 inhibitors, increasing the level of statins, hence causing the toxicity3. Clinical studies demonstrated that verapamil and simvastatin coadministration increases the latter’s concentration by fivefold4. Besides increased concentration of pro-drug, metabolite and half-life, multiple pathways contribute to myofiber necrosis, via mitochondria-mediated apoptosis, myocyte cellular membrane instability, and reduced coenzyme affecting mitochondrial function 2, 5, 6. Other risk factors include renal insufficiency, hypothyroidism, hepatic dysfunction, macrolide and antifungals7, 8. Our patient had risk factors including CKD, hypothyroidism, and verapamil use. Per an FDA safety alert in 2010 the daily dose of simvastatin should be limited to 20mg, when co-administered with verapamil. This unique case presents multiple learning opportunities for internal medicine physicians. It stresses the importance of medication review and cross-checking drug-drug interactions, that are frequently overlooked in patients with hypertension and hyperlipidemia. Additionally, a comprehensive history and medication reconciliation spared the patient from unnecessary tests and prolonged hospitalization as the only definitive treatment is discontinuation of offending agents and supportive care.

References


INDIANA CLINICAL VIGNETTE POSTER FINALIST - JENNIFER L PENG, MD

Two Roads Diverged in a Yellow Patient

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Introduction: Pancreatic tuberculosis is a rare clinical diagnosis as its presentation usually mimics pancreatic cancer.1 We report the case of a patient with a pancreatic mass found to be a tuberculoma on pathological examination.

Case Presentation: Our patient is a 75-year-old male leather worker from Mexico who was brought to the ED by his daughter-in-law with new-onset, moderate-to-severe epigastric and left upper quadrant abdominal pain as well as nausea with non-bilious, non-bloody emesis for 5 days. He also reported subjective fever, chills, and increased diaphoresis over a few days. He had unintentional three-kilogram (kg) weight loss over the preceding three months. He denied any abdominal distention, constipation, diarrhea, hematochezia, or melena. He denied recent dyspnea, cough, oral ulcers, arthralgia, or back pain. Physical examination was significant for tachycardia, jaundice, and a soft abdomen with a directly tender epigastrium but no rebound. There was no flank fullness, shifting dullness, or stigmata of chronic liver disease. Laboratory testing revealed leukocytosis with bandemia, hyperbilirubinemia (7.4 mg/dL [direct 5.6 mg/dL]), elevated alkaline phosphatase, and hypoalbuminemia. Further testing showed elevated lactate dehydrogenase, lipase, CA19-9 (1613 U/mL), and CA125 (4101 U/mL). Enhanced CT of the abdomen demonstrated findings concerning for an ampullary or pancreatic mass causing diffuse biliary and pancreatic ductal dilatation. There also was a right hepatic lobe lesion and periportal/peripancreatic adenopathy concerning for metastatic disease with complete occlusion of the left portal vein. The patient was started on piperacillin/tazobactam for possible cholangitis on admission. On hospital day 4, the patient underwent endoscopic ultrasound (EUS) that showed a mass of the pancreatic uncinate process with diffuse pancreatic and biliary ductal dilatation in addition to peripancreatic, porta hepatitis, and retroperitoneal lymphadenopathy. On the same day, he underwent endoscopic retrograde cholangiopancreatography (ERCP) with stent placement and fine needle aspiration (FNA). Pancreatic FNA was inconclusive and showed reactive lymphoid tissue. Hospital course was further complicated by thrombocytopenia and anemia requiring numerous transfusions which led to a bone marrow biopsy showing hypercellular marrow with maturing trilineage hematopoiesis. Ultimately, infectious work-up was significant for a positive T-spot assay. A repeat pancreatic biopsy was completed, and the pancreatic mass was found to be a tuberculoma on pathological examination.

Discussion: Pancreatic tuberculosis can have myriad presentations and imaging studies usually show pancreatic masses, cystic lesions, or abscesses.2,3 Pancreatic tuberculosis should be considered in patients presenting with pancreatic lesions and peripancreatic lymph nodes. The most appropriate diagnostic tools include CT of the abdomen and EUS-guided biopsy or FNA. Complete cure can be achieved with standard anti-tuberculosis therapy.4 Of note, the elevation in CA19-9 to ~1600 U/mL is more consistent with malignant neoplasms of the pancreaticobiliary system although such elevations have been reported in benign diseases of the pancreas as well.5

References

An Unusual Presentation of Amyopathic Dermatomyositis with Interstitial Lung Disease

Authors: Russell Purpura M.D., Mrisa Sahai M.D., Rajat Kapoor M.D.

Introduction: Subcutaneous emphysema (SE) is an unusual skin finding that is often clinically striking. This finding is most commonly seen in pneumothorax cases from chronic obstructive pulmonary disease, trauma or iatrogenic. SE is typically seen on the chest and neck, while treatment is aimed at the underlying pathology. However, SE secondary to an inflammatory myopathy disease is highly unusual.

Case Presentation: A 60 year old female presented to the emergency department for dyspnea and facial swelling for two weeks. There was a dark purple-red rash on her hands, chest and around her eyes. She denied fevers or myalgias and endorsed a twenty-pound weight loss over two months. Her pertinent vital signs include oxygen saturation of 86% on room air that corrected to 97% with two liters of oxygen. Physical exam showed extensive crepitus on palpation throughout the chest, neck and face, along with crackles in all lung fields on auscultation. Skin exam demonstrated a periocular red-purple (“heliotrope”) eruption, violaceous rash distributed on chest and Gottron’s papules most notably on the metacarpal phalangeal joints with periungual erythema around the nail beds.

CT Imaging notable for severe pneumomediastinum and subcutaneous emphysema in neck and face that extends to zygomatic arches on the left. Lung fields demonstrated ground glass opacities and consolidation in the lung bases. Extensive lab work up showed normal creatine kinase, aldolase, erythrocyte sedimentation rate, negative anti-jo 1, rheumatoid factor, and myomarker panel 3. She was started on high dose prednisone and was discharged with two liters of oxygen with close follow up with rheumatology and pulmonology.

A bronchoscopy with lavage (BAL) and transbronchial biopsy was performed as outpatient. The BAL was suggestive of cryptogenic-organizing pneumonitis (COP) and pathology reported interstitial fibrosis. At the next encounter, the patient was diagnosed with amyopathic dermatomyositis (ADM) with associated interstitial lung disease (ILD) based on th clinical findings. Mycophenolate mofetil was added to her regimen, slow prednisone taper was started and she was maintained on two liters of oxygen. Follow up imaging six months later showed resolution of neck and facial SE, but residual pneumomediastinum was still present.

Discussion: ADM is a rare autoimmune condition in which patients present with pathognomonic skin findings associated with dermatomyositis, but lack the inflammatory myopathy component. ADM has a high prevalence of ILD ranging from 30-50%. ADM-associated ILD has a high mortality up to 55% within six months and even worse prognosis if associated with SE and pneumomediastinum. This case is unique in that it presented with extensive cutaneous rash, SE and ILD findings all at the initial encounter. Literature review has only reported a handful of similar cases with all three findings. Fortunately for our patient, she responded well to prednisone, mycophenolate mofetil and did not require uptitration of oxygen.

References

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Non-Uremic Calciphylaxis: Exploring Two Rare Associations

Authors: Cory Powers, MD, Sam Lampe, DO, Sarah Seyffert, MD, Antoine Saliba, MD

Introduction: Non-uremic calciphylaxis has been rarely reported in patients with alcoholic liver cirrhosis or gastric bypass surgery. Insufficient attention has been given to the adverse effects associated with the therapies used to supplement expected deficiencies, especially in patients with other risk factors for rare but potentially fatal disease, calciphylaxis. We present a case of nonuremic calciphylaxis in a normocalcemic obese female patient with alcoholic liver cirrhosis. We discuss how the different comorbidities our patient had could have contributed to the pathogenesis of calciphylaxis.

Case Presentation: Our patient is a 29-year-old White woman, with a past medical history significant for recently diagnosed alcoholic cirrhosis, proven by liver biopsy, and Roux-en-Y gastric bypass surgery, who was admitted to the intensive care unit for hepatic encephalopathy and upper gastrointestinal bleeding. She had been discharged from the hospital five days prior with an initial presentation of a painful, erythematous rash on her lower extremities. On physical exam, the rash had dark erythematous borders and a necrotic center with associated bullae and desquamation. Superficial biopsy of the rash was concerning for thrombotic vasculopathy. Laboratory findings were notable for: serum urea nitrogen 23 mg/dL (eventually improved to 8 mg/dL), serum creatinine 2.87 mg/dL (eventually improved to 0.63 mg/dL), calcium 8.8 mg/dL, phosphorus 4.7 mg/dL, parathyroid hormone 46 pg/mL, and albumin 3.0 g/dL. She was found to have low protein C activity 22% and was started on apixaban during her first hospital stay. Apixaban was stopped after her presentation with an upper gastrointestinal hemorrhage. Hematologic workup was negative for consumptive coagulopathy. Repeat protein C activity was found to be 44%. On deep tissue biopsy, the subcutaneous fat showed multiple thin-walled medium-sized blood vessels with calcification and scattered foci of soft tissue calcification. Von Kossa special stain confirmed findings consistent with calciphylaxis. Treatment was started with wound therapy and sodium thiosulfate intravenously three times weekly. The patient was discharged thereafter to seek further medical care at another medical center. She was advised to continue sodium thiosulfate therapy and wound therapy.

Discussion: The most commonly identified conditions associated with non-uremic calciphylaxis include hyperparathyroidism, malignancy, and alcoholic liver disease. Previous corticosteroid therapy, albumin or blood transfusions, and protein C and S deficiencies have been described in patients with nonuremic calciphylaxis. The patient discussed in this case report did not have ESRD. Her 24-hour creatinine clearance estimated her glomerular filtration rate to be 120 mL/min. Our patient was treated with prednisone for alcoholic hepatitis two months prior to presentation and had received blood products during a previous hospital stay. Identification of risk factors for calciphylaxis and careful analysis of risks and benefits of therapeutic interventions are necessary in patients who are morbidly obese and have undergone bariatric surgery to avoid this potentially fatal disease and its repercussions.
A rare case of adult RSV croup, a restricting diagnosis.

Authors: Alexander Slaten, DO; Azmina Alibhai, MD; Brandon Perkins, DO

Introduction: Croup or laryngotraheitis, most commonly from parainfluenza-1 and respiratory syncytial virus (RSV), is a common pediatric illness that varies in severity. RSV is a virus that can affect all ages; however, in adults most infections with RSV are mild. We present a severe case of adult laryngotracheobronchitis due to RSV, which required intensive care and was successfully managed with mechanical ventilation and corticosteroids.

Case Presentation: A 71-year-old female presented to the emergency department (ED) with three days of dry cough and worsening dyspnea. Past medical history was significant for diastolic dysfunction and mild pulmonary hypertension. On arrival, she was in hypoxic respiratory distress with inspiratory stridor and tachycardia. She was initially managed with racemic epinephrine, Benadryl, and Pepcid for a possible anaphylactic reaction. She was subsequently intubated after no improvement in her condition. Direct laryngoscopy revealed mildly edematous vocal cords. Chest x-ray was unremarkable. CTA revealed marked subglottic tracheal narrowing with extension into left mainstem bronchus. The images were reviewed by thoracic surgery that felt the stenosis was chronic and not contributing to her acute illness. She was empirically treated with antibiotics for community-acquired pneumonia while causes of tracheobronchial stenosis were being investigated. This included extrinsic causes, tracheomalacia, chronic inflammatory and infiltrative conditions such as relapsing polychondritis, Granulomatosis with polyangitis, rheumatoid arthritis, amyloidosis, and chronic gastroesophageal reflux. Besides a mildly elevated ESR at 37, ANCA, ANA, and RF were negative and no prior iatrogenic causes were discovered. She subsequently developed fevers and respiratory panel returned positive for RSV. After two days of aggressive management with corticosteroids, high resolution CT revealed improvement in tracheal narrowing. She was successfully extubated on day six of admission and discharged with complete recovery of her respiratory function.

Discussion: Respiratory syncytial virus is a common cause of croup-like illness resulting in laryngeal and tracheal inflammation in the pediatric population, often requiring hospitalization. Healthy younger adults infected with RSV typically have symptoms restricted to the upper respiratory tract. However, RSV is a significantly under recognized cause of severe lower respiratory complications in older adults, resulting in respiratory failure and mortality similar to influenza in the winter months. To our knowledge, very few documented cases exist of adults with laryngotracheobronchitis secondary to RSV. Thus, guidelines for management are unavailable. Per literature review, adults have been successfully treated with various combinations of nebulized racemic epinephrine, corticosteroids, and helium-oxygen mixture. Our patient did not show improvement with nebulized epinephrine, but responded adequately to corticosteroids.

Our case reveals a rare incident of severe adult laryngotracheobronchitis as a result of respiratory syncytial virus. It highlights the necessity to remain astute to broad etiologies of acute respiratory failure in older adults. Early recognition and aggressive management of RSV in older adults is a necessity to reduce mortality.
Acute abdomen and crushing chest pain following surgery

Authors: Jason A. Cascio, MD, PhD, Mentor: Jeydith Gutierrez Perez, MD

Introduction: Ms. S is a 60yo female with a history of Familial Mediterranean Fever (FMF) who presented for further evaluation of persistent tenosynovitis of the right middle finger, which developed after trigger finger release surgery months before. Two subsequent incision and drainage (I&D) procedures were done at a previous facility without improvement. On admission, she was afebrile and complained of severe right hand pain. Cultures from previous I&D grew Pseudomonas, E. faecalis, and S. mitis. She was treated with IV daptomycin and cefepime for several days but had worsening pain and swelling of the hand. Thus, she underwent a third surgical I&D by the orthopedic hand service.

Case Presentation: On post-op day 0, Ms. S. developed sudden onset, 10/10 generalized abdominal pain with fever to 39.0 C. Exam was significant for rigid abdomen with guarding and rebound tenderness. She was given oral prednisone and IV morphine, with improvement in her pain and fever. The next morning, a rapid response was called. On arrival, patient was in distress and diaphoretic, complaining of 10/10 crushing, retrosternal chest pain and difficulty breathing. She was afebrile, but tachycardic to the 130s, hypertensive to 200/130, and SpO2 87% on room air. Stat EKG and troponin were normal and point of care ultrasound was reassuring. She was again treated with oral prednisone and IV morphine, as well as sublingual nitroglycerin and IV metoprolol with improvement in her symptoms. Troponin remained negative x 3. She was transitioned to IV morphine PCA and given 3 more days of oral steroids. Her pain quickly subsided. She was discharged 4 days later pain free with a PICC line in place for continued IV antibiotics. Her post-op presentation of an apparent surgical abdomen and crushing chest pain was consistent with a FMF flare.

Discussion: FMF is an autoinflammatory disorder characterized by sporadic, recurrent bouts of fever and serosal inflammation, typically manifesting as abdominal pain or chest pain. Vigorous exercise, emotional stress, or surgeries are common triggers, but often none can be identified. Treatment is with colchicine, which prevents acute attacks, subclinical inflammation between attacks, and slows progression toward long term complications such as secondary amyloidosis. Diagnosis should be considered in patients from at risk ethnic groups with recurrent febrile episodes accompanied by peritonitis, synovitis, or pleuritis. For patients carrying a diagnosis of FMF, knowledge of the typical symptoms and triggers can guide a judicious workup in the event of an acute flare.

References

Rapid Progressive Dementia with Sporadic Creutzfeldt-Jakob Disease

Authors: Sinan Khayyat, MD, Daly Al-Hadeethi, MD, Mohinder R. Vindhyal, M.D., M.Ed., Kansas University – School of Medicine -Wichita / Department of Internal Medicine

Introduction: Prion diseases are rare neurodegenerative diseases which usually have long incubation periods and rapidly worsening neurological outcomes. Creutzfeldt-Jakob disease (CJD) is a rare human prion disease with progressive, fatal encephalopathy characterized by dementia, cerebellar ataxia, and visual disturbances. CJD is caused by an abnormal protease-resistant isoform of prion protein (PrPSc), a misfolded version of the normal cellular isoform (PrPC)\textsuperscript{1,2}. It is a rare disease with an incidence rate of approximately 1 case per million population per year with a worldwide distribution. We are presenting an 85-years-old male with Creutzfeldt-Jakob disease (CJD).

Case Presentation: An 85-year-old man presented with confusion, aphasia, and episodic seizure-like activity in his right upper extremity and right eye twitching for two weeks duration. Neurological exam revealed stupor, aphasia, verbally non-responsive, generalized weakness, and lower extremity myoclonic movements. Pupils were constricted but reactive to light with no facial asymmetry. Cogwheel rigidity was noticed with only equivocal left Babinski. Workup for confusion showed generalized cerebral atrophy and chronic microvascular changes with scattered focal and confluent areas of nonspecific T2 bright signal in the periventricular and subcortical white matter but no acute events on MRI. EEG showed diffuse slowing with transient generalized epileptiform discharges for which the patient was started on anti-seizure medication. Lumbar puncture ruled out meningitis/encephalitis. CSF studies were negative for infectious and parasitic etiology. The CSF studies were also sent for 14-3-3 antigen test, which was elevated, Tau protein level was >4000, and Real-Time Quaking-Induced Conversion assay was also positive. The clinical diagnosis of CJD was given. The patient was discharged to hospice and passed away soon after that. An autopsy was done after and results were evident for sporadic CJD with MM1 gene sequence.

Discussion: The definitive diagnosis of CJD is made with a brain tissue biopsy and western blot confirmation of protease-resistant PrP and presence of scrapie-associated fibrils. Criteria for the diagnosis of probable sporadic CJD in patients presenting with rapid progressive dementia are 2 of the following: myoclonus, visual or cerebellar signs, pyramidal/extrapyramidal signs, akinetic mutism, and a positive result of either a typical EEG with periodic sharp wave complexes and/or a positive 14-3-3 (CSF) assay. Our patient met criteria of probable sporadic CJD with the presence of progressive dementia, akinetic mutism, myoclonus, and an extrapyramidal sign of rigidity with positive 14-3-3 CSF assay. The patient also tested positive on the RT-QuIC assay which has highly specific for CJD disease and diagnosis later confirmed with an autopsy of brain tissue and further genetic testing. Prognosis with CJD has a rapid, deteriorating course invariably, with death generally within two years of symptom onset. No treatment has been proposed so far, and few medications help with symptoms. Early diagnosis also allows patients and their families to prepare for the expected disease course.

References

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KANSAS CLINICAL VIGNETTE POSTER FINALIST - ERIK A CALDERON, MD

Extensive Deep Venous Thrombosis in a 31-Year-Old Female on Oral Contraceptives Diagnosed with May-Thurner Syndrome

Authors: Erik Calderon, MD, Brent Duran, DO, Stephanie Pankow, DO

Introduction: May-Thurner Syndrome is a rare but important diagnosis in a small percentage of new deep venous thrombosis diagnoses. The prevalence is unknown but approximated to be 2-5% of the causes of symptomatic deep venous thromboses. Risk factors include female gender, post-partum, and oral contraceptives. It is essential that providers are aware of the high rate of recurrence in patients with this diagnosis and act to decrease other modifiable risk factors such as oral contraceptives.

Case Presentation: A 31-year-old female, who was 3 months post-partum following a C-section, presented to the hospital with left lower extremity pain and swelling of one-day duration. She had started oral contraceptives 3 weeks prior to presentation. On physical exam, she had left lower extremity edema and palpable dorsalis pedis pulses. There was no warmth or tenderness to palpation. Ultrasound showed extensive thrombus throughout the left lower extremity venous system involving the popliteal to the common femoral veins with extension into the external iliac vein. There was no personal or family history of hypercoagulability. She was given a dose of therapeutic enoxaparin at that time and admitted for further management. A heparin drip was initiated while bridging to warfarin. Due to the extensive nature of her thrombus, she received catheter-directed TPA treatment to her left lower extremity and was found to have a chronically occluded inferior vena cava with a large clot burden. At that time, she was diagnosed with May-Thurner Syndrome. The patient decided to leave before bridging was complete. She left the hospital on therapeutic enoxaparin with arrangements to have outpatient stent placement performed by interventional radiology. Upon discharge, the oral contraceptives were discontinued and lower extremity stockings were placed. She likely will require long-term anticoagulation to prevent any further thrombus formations.

Discussion: May-Thurner Syndrome, also known as iliac vein compression syndrome, is an extrinsic venous compression by the arterial system against bony structures in the iliocaval venous territory. The most common variant occurs when the left iliac vein is compressed between the right common iliac artery and the L5 vertebrae. Many times, a patient will be asymptomatic when there is only a partial obstruction, but it can progress to more symptomatic extensive deep vein thrombosis as was seen in this patient. Although uncommon, it is important to differentiate May-Thurner Syndrome as a cause of deep venous thrombosis as the treatment modalities and long term anticoagulation may differ.
Polymorphous low-grade neuroepithelial tumor of the young: A distinct biological entity with excellent surgical prognosis

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Introduction: Polymorphous low-grade neuroepithelial tumor of the young (PLNTY) is a recently characterized subset of neuroepithelial neoplasms, of which exceedingly few cases have been reported in the literature to date. These tumors appear to demonstrate excellent definitive response to surgical resection, underscoring the importance of recognizing them amidst the fuller spectrum of low-grade neuroepithelial tumors.

Case Presentation: A 15-year-old previously healthy female presented for evaluation of new onset grand mal seizures. MRI brain with contrast months before and on the day of surgery revealed a solid and cystic superficial left posterior temporal lobe mass without definitive evidence of enhancement. Neurosurgery was consulted and recommended surgical excision of the mass and the patient experienced full resolution of seizure activity postoperatively. Histopathologic analysis of the resected specimen showed extensive microcalcifications with tumor cell populations consisting of round uniform nuclei and clear cytoplasm resembling oligodendrial cells. Tumor cellularity was moderate without definite evidence of mitotic activity, microvascular proliferation, or necrosis. The cells appeared to have a complex phenotype showing expression of Olig2 and CD34 (tumor cells) as well as synaptophysin (tumor cells and parenchyma) suggestive of both glial and neuronal differentiation. Immunohistochemical staining for GFAP was diffusely positive, largely corresponding to an underlying pattern of reactive gliosis. Notably, no BRAF mutation was identified including BRAFV600E, based on the neurooncology targeted next-generation sequencing (NGS) panel. Mutant IDH1 (IDH1-R132H) was also negative, however other less common IDH1/IDH2 mutations were not entirely excluded. Finally, ATRX expression was attained in tumor cells and P53 protein was variably expressed in a moderate number of cells. In light of the above, the tumor was most histopathologically consistent with PLNTY, recently reported (Acta Neuropath 201;133:417-29). Postoperative outpatient follow-up demonstrated persistent cessation of convulsive seizures for the patient in question several months out from treatment.

Discussion: PLNTYs are a newly characterized distinct subgroup of low-grade neuroepithelial tumors possessing infiltrative growth, oligodendroglioma-like cellular components, and intense CD34 positivity. Though capable of inducing convulsive seizures, surgical excision favors prompt and sustained postoperative resolution of epileptogenic activity, further accentuating the importance of recognizing this new subclass of rare neoplasms.

References

Prosthetic joint infection secondary to Mycobacterium avium complex presenting as FUO

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Introduction: Non-tuberculous mycobacterium (NTM) are commonly pathogenic in the HIV/AIDS population and in patients with chronic lung disease such as cystic fibrosis and bronchiectasis. NTM are ubiquitous in the environment, in soil and water worldwide. Mycobacterium avium complex (MAC) is the most common NTM infection in the United States, with many asymptomatic carriers. Those who have symptomatic infection generally present with progressive lung disease, characterized by bronchiectasis with patchy interstitial infiltrates and cavities. Localized disease is rarely observed, as it was in our patient.

Case Presentation: A 56-year-old male with rheumatoid arthritis on abatacept, hydroxychloroquine and sulfasalazine presented to clinic with a 3-month history of malaise and low-grade fever. He returned from a mission trip to Honduras 3 months previously. He developed low-grade fever a month later, with unintentional weight loss and night sweats. White blood cell count was 6.3 with hemoglobin of 12.4 grams, chest radiograph was normal. Two sets of blood cultures were negative and echocardiogram was normal. He developed progressive right hip pain (in the setting of remote right hip arthroplasty). A temperature of 103 degrees prompted admission. He was monogamous, denied tobacco or recreational drug use. He drank only bottled water in Honduras with no adventurous eating. He had no other travel or animal exposures. On examination, extremities were atraumatic without synovitis; and decreased active range of motion of the right hip. ESR was 25. An MRI of the right hip joint showed fluid collections medial to the lesser trochanter. Joint aspirate was grossly purulent, thus the patient was taken to the operating room for a prosthetic explant with antibiotic spacer placement. Culture grew acid-fast bacteria at 10 days, identified as MAC. Initial broad-spectrum antibiotics were adjusted and he was discharged on intravenous amikacin three times weekly with oral rifampin, ethambutol, and azithromycin. Amikacin was discontinued after 4 weeks of treatment. The remainder of therapy was continued for 12 months. His immunosuppression for rheumatoid arthritis was lowered. Eleven months after treatment initiation, repeat joint aspiration culture was negative. He underwent repeat right hip arthroplasty and tissue cultures were negative.

Discussion: NTM prosthetic joint infections are a rare presentation of NTM infection. Few case reports of MAC prosthetic joint infection exist in the literature. Most occurred in post-transplant (renal and cardiac transplant) patients [3]. One occurred in a patient on chronic steroids for rheumatoid arthritis [4]. Future increased use of biologics and other immune-modulating therapy in rheumatoid disease and malignancy the will increase the incidence of NTM infection. Optimal duration of therapy for non-pulmonary NTM is unclear, but guidelines for pulmonary NTM recommend 6-12 months of treatment [2]. Investigation on the appropriate choice, interval, and duration of treatment for NTM prosthetic joint infections is imperative.

References

Gastrointestinal bleeding in AL Amyloidosis

Authors: Samuel B. Reynolds, MD, Harrison Daniel, MD, Khushboo Gala, MD, NiharShah, MD

Introduction: Acute gastrointestinal bleeding has a wide differential, and often requires endoscopy to detect a source. Comorbid plasma cell dyscrasias, particularly when patients are undergoing stem cell transplant, makes diagnosis and subsequent management even more challenging.

Case Presentation: A 70-year-old Caucasian man admitted to the hospital with a hemoglobin of 7.5 and stable vital signs. History of systemic AL amyloidosis, treated with 4 cycles of combination dexamethasone, ixazomib and lenalidomide, followed by autologous stem cell transplant. Approximately a month post-transplant, the patient developed second episode of hematochezia; colonoscopy 8 months previously for similar complaint demonstrated internal and external hemorrhoids, with ulcerations of the colonic mucosa. He was also noticed to have multiple amyloid deposits in his oral cavity with accompanying macroglossia. He had recurrent hematochezia, after which he became hypotensive and unresponsive, requiring intubation. Repeat colonoscopy demonstrated a visible blood vessel emerging from a 3 cm ulcer of the ascending colon with clean margins as well as a circumferential, clean-based ulcer of the transverse colon. A hemoclip was applied to the ascending colon ulcer. The patient was extubated and was discharged 8 days after admission without any further bleeding. Approximately one year later, he remains under maintenance therapy, without recurrent bleeding.

Discussion: Gastrointestinal involvement in systemic amyloidosis is reported in 79% of cases, with deposits being found anywhere along the GI tract, although, in order, the duodenum, stomach, and rectum are most commonly involved [4,9]. Oral lesions can occur in both AA and AL amyloidosis, but tend to be more common in AL, and are themselves rare, as oral amyloid more commonly presents as macroglossia [11]. Regarding the pathophysiology of GI amyloidosis, increased frailty of the adjacent blood vessels results in local ischemia, infarction, and mucosal injury [4,8]. AL and AA amyloidosis are, respectively, associated with bulk deposition causing mucosal protrusions and diffuse deposition causing mucosal friability and ulcerations.

Regarding therapy, there are no definite recommendations for the management of bleeding and gastrointestinal involvement other than conventional supportive and symptomatic therapy, but usually the course of the disease is relatively benign [2,3]. Guidelines indicate that endoscopic management within 24 hours of bleeding leads to better outcomes [5]. This concept is illustrated in our patient, who, despite negative endoscopies on initial presentation, was discovered to have a source bleeding, requiring lifesaving intervention [6,7].

One challenge in interpreting this case is that biopsy was not performed, so it cannot be said definitively that the bleeding was due to amyloid deposition in the colon. This, however, illustrates an important teaching point in maintaining an open different when approaching GI bleeding in a patient with established systemic amyloidosis: it is important for providers to consider other reversible causes (e.g. ischemia, as in this patient) when approaching patients.

References

KENTUCKY CLINICAL VIGNETTE POSTER FINALIST - EMMANUEL OKENYE, DO

MALT on the Lips

Authors: Emmanuel Okenye, DO; Avinash Aravantagi, MD; Shumar Shekah, MD

Introduction: Mucosa Associated Lymphoid Tissue (MALT) are a subtype of marginal zone lymphoma which arise from post germinal center marginal zone B cells. These along with Non Hodgkin’s lymphoma share similar immunophenotype - positive CD19, CD20, CD22 and negative CD5, CD10 and CD23. It is often associated with Helicobacter pylori infection of the GI tract. Although they can affect other mucosal sites including the lips, they have been very seldom documented in medical literature. We present a case of a rarely occurring MALT lymphoma of the lower lip.

Case Presentation: A healthy 45 year old Caucasian female presented to the dentist’s office with lower lip swelling for 6 months. She had no dental caries. She was initially treated with antibiotics for possible infected mucocele. Swelling was about 10mm x 8mm. It was tender to palpate, and progressively worsened. She was referred to oral maxillofacial surgery. She had no history of radiation exposure or prior cosmetic procedures to lip. No history of travel or animal bites.

Her lab work which included CBC, CMP and CRP were normal. She had excision biopsy of the lower lip swelling by oral maxillofacial surgeon and histology showed marginal B cell lymphoma- MALT lymphoma. She was evaluated by Oncology, then had CT Neck, CT abdomen & pelvis, and PET CT to rule out any disseminated disease. These imaging studies were unremarkable. An Esophagoduodenoscopy and a Colonoscopy didn’t reveal any mucosal abnormality or mass. Gastric biopsy was negative for Helicobacter Pylori negative gastritis.

After it was confirmed as an isolated Non Hodgkin’s Lymphoma (NHL); a primary extra-nodal variant with no dissemination elsewhere in the body, it was decided to pursue a radiation treatment course and not instigate any further treatment, taking into consideration the patient’s quality of life and psychological well being. The patient has been kept under close observation and clinically, patient is in good health.

Discussion: In this case, non-tender swelling in lower lips, which mimicked an infected mucocele, would have been probably misdiagnosed if a thorough systemic evaluation was not done. Therefore, any simple asymptomatic lesion or swelling in the labial mucosal region should be screened for underlying systemic diseases and biopsied, and then Immunohistochemistry plays a final vital role in tailoring treatment.

References

HINTS to a Diagnosis of Multiple Sclerosis

Authors: Devin Bourgeois, MD, Robert Thibodaux, MD- Internal Medicine Residency Program, LSUHSC-Baton Rouge

Introduction: Vertigo is the illusion of movement usually accompanied by nystagmus and distressing symptoms such as nausea, vomiting, pallor and postural instability. It is categorized as either peripheral or central, the latter being associated with significant morbidity. This case illustrates the importance of distinguishing central versus peripheral etiologies of vertigo which ultimately led to a diagnosis of multiple sclerosis (MS).

Case Presentation: A 30-year-old female with hypertension, scoliosis, and obesity presented with several hours of acute vertigo, nausea and vomiting. She noted a brief episode of vertigo with N/V earlier in the day with sitting up but denied any headache, changes in vision, weakness or numbness, hearing loss, tinnitus, or recent URI. She was evaluated a few months prior by ENT for decreased hearing and right ear fullness and diagnosed with eustachian tube dysfunction. Around that time, she underwent PT for LLE weakness and numbness which was attributed to her scoliosis. Exam revealed no focal motor or sensory deficits. HINTS(head impulse, gaze-changing nystagmus, test of skew) exam revealed normal head impulse and gaze changing nystagmus consistent with central vestibular dysfunction. Subsequent MRI of the brain, cervical, and thoracic regions suggested multiple foci of active demyelination and a lumbar puncture revealed >5 oligoclonal bands in CSF, both consistent with MS. Labs including RPR, HIV, ANA, ESR, CRP, B-12, and complements were unrevealing. After 3 daily doses of pulse dose steroids and PT, she was significantly improved and discharged.

Discussion: MS is an autoimmune inflammatory disorder of the CNS affecting the optic nerve, brain, and spinal cord. While vertigo symptoms are noted in up to 50% of patients with MS at some point during the disease, the etiology is usually peripheral with BPPV being very prevalent. However, a central etiology of vertigo due to demyelination in vestibular pathways is the presenting symptom in only 5% of cases. HINTS exam is a simple bedside test which is excellent at distinguishing central from peripheral vestibular dysfunction. The presence of either a normal head impulse, gaze-changing nystagmus or skew deviation suggests a central etiology. In this case, results of the HINTS exam along with her prior symptoms of LLE numbness and weakness was concerning for CNS pathology, namely MS. As a result, she underwent proper neuroimaging as well as LP which allowed her to be properly diagnosed with MS.
RARE PRESENTATION OF CLOSTRIDIUM DIFFICILE BACTEREMIA

Introduction: *Clostridium difficile* is a spore forming gram-positive anaerobic rod, often associated with diarrhea and pseudomembranous colitis. *C. difficile* infections are limited to the colon and there are very few cases of extra intestinal *C. difficile* infections (Ex-CDI) such as *C. difficile* bacteremia (CDB), osteomyelitis, visceral abscess, empyema, etc. CDB is more frequently reported than other Ex-CDI. CDB is a polymicrobial disease and the treatment is directed against all microorganisms. We describe a case of CDB who presented initially with appendicitis.

Case Presentation: A 77-year-old male presented with epigastric pain, nausea and vomiting. Physical examination revealed tenderness and guarding in the right lower quadrant of the abdomen. A CT scan of the abdomen and pelvis reported acute appendicitis and patient underwent urgent laparoscopic appendectomy. Intraoperative findings were significant for ruptured appendix and he was thus started on antibiotic therapy with intravenous piperacillin-tazobactam.

Blood cultures obtained on the day of admission grew *Escherichia coli* and a gram-positive rod later identified as *Clostridium difficile*. Four days later, he had several small loose bowel movements, *C. difficile* PCR was positive and blood cultures grew *C. difficile* in addition to *Eggerthella lenta*. His antibiotic regimen was changed to ceftriaxone and metronidazole. On day 6, blood cultures became sterile. On Day 7, CT of the abdomen and pelvis showed an abscess in the right lower quadrant, which was drained under CT guidance and a drain was placed in the abscess. Culture of the abscess grew *Candida albicans* only. He was discharged on oral metronidazole and fluconazole.

Discussion: The incidence of Ex-CDI is approximately 0.17% with mortality rate up to 36% depending on co-morbidities. CDB patients usually do not present with diarrhea/colitis symptoms on admission and recent antibiotic use is rarely present. When GI pathology is present, normal commensals of gut such as *C. difficile* can translocate to blood and cause bacteremia. CDB is usually occurs along with other GI flora (polymicrobial) in the blood. Polymicrobial and monomicrobial CDB has higher mortality rate since *Clostridium* species have been shown to enhance the pathogenicity of other organisms in polymicrobial CDB. Our case is a polymicrobial CDB associated with appendicitis and colonic abscess. However, the abscess fluid did not grow *C. difficile* likely due to antibiotic administration prior to aspiration.

References


A case of Lymphomatosis Cerebri presenting as rhombencephalitis

Authors: Rebecca P. Bystrom, MD and Christine Lu-Emerson, MD, Departments of Medicine and Neurology, Maine Medical Center, Portland ME

Introduction: Lymphomatosis Cerebri is a rare variant of primary CNS lymphoma (PCNSL) with less than 50 reported cases. Its clinical and radiographic presentation may be nonspecific, unlike classic PCNSL. This enables it to masquerade as non-malignant infectious or inflammatory conditions, resulting in diagnostic delay.

Case Presentation: We present a 62-year-old woman with minimal past medical history, limited only to tobacco and marijuana use, who transferred to our facility with rhombencephalitis. Twelve days prior to admission she developed vertigo, diplopia and upper respiratory infectious symptoms. She presented to an outpatient clinic with subjective fevers, chills, diaphoresis, and photophobia, and was treated for an acute exacerbation of presumed COPD. Seven days later she experienced dysphagia, right-sided facial droop, drooling, and word-finding difficulty following a fall. She was admitted to her local hospital. In retrospect, she had several months of fatigue, ataxia, new snoring, hearing loss, and memory impairment. Her initial MRI showed confluent T2 hyperintensity within the pons, midbrain, and left basal ganglia.

On transfer to our facility, the patient was tachypneic but afebrile. Physical exam was significant for mild respiratory distress, diffuse rhonchi, and occasional end-expiratory wheeze. The neurological exam showed mild right-sided facial droop, nystagmus, a non-abducting right eye, dysarthric and gravelly speech, and poor coordination of the right upper extremity. There was mild peripheral leukocytosis as well as a mixed chronic respiratory alkalosis and metabolic acidosis. Lumbar puncture showed CSF pleocytosis with lymphocytic predominance and normal protein and glucose. Non-contrast CT of the chest, abdomen, and pelvis was unremarkable.

Our patient had a protracted hospital course extending over 80 days. The broad differential diagnosis for rhombencephalitis included infectious, inflammatory, and demyelinating etiologies. Over the course of the admission three lumbar punctures showed no evidence of malignancy on CSF cytology or flow cytometry. The patient underwent two rounds of pulse steroids and plasmapheresis, with limited clinical and radiographic improvement, and ultimately had clinical deterioration. Subsequent MRIs showed progression of the T2 hyperintensity into the thalamus, bilateral basal ganglia, and periventricular white matter. There was new heterogeneous enhancement and restricted diffusion. Ultimately, a new lesion developed in the right frontal lobe and a brain biopsy showed diffuse large B-cell lymphoma.

Discussion: Lymphomatosis Cerebri, unlike classic PCNSL, presents with nonspecific diffuse intracranial lesions that are often poorly circumscribed with absent or minimal contrast enhancement. There may be variable restricted diffusion, and absent or minimal mass effect. These vague characteristics are often nonsuggestive of malignancy and result in delayed diagnosis. This case demonstrates that Lymphomatosis Cerebri can be a great “mimicker” of other diseases, highlighting the importance of tissue diagnosis, even in the setting of multiple negative CSF studies.

References

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Diffuse large B-cell lymphoma masquerading as hepatocellular carcinoma in a patient with alcoholic cirrhosis.

Authors: Meng, Z., Sharma, A.

Introduction: Hepatic lesions in the cirrhotic liver can be a diagnostic dilemma as the differential diagnosis can be quite broad including numerous benign and malignant conditions. Of the malignant entities, hepatocellular carcinoma, hepatic lymphoma and metastases are causes that are important to differentiate as the treatment modalities and prognosis would be substantially different.

Case Presentation: A 66 year-old male with a 20 year history of alcoholic cirrhosis presented to his family practitioner with a one-month history of confusion, day-night reversal, aggression, generalized weakness, constipation, 15-pound weight loss and worsening peripheral edema. Urgent liver ultrasound showed multiple hypoechoic hepatic lesions that were not present during his last ultrasound 6 months ago concerning for multifocal hepatocellular carcinoma (HCC).

Initial physical exam revealed a nodular liver edge palpable 1.5 cm below the right costal margin and pitting edema extending up to mid-shin bilaterally. Examination was otherwise unremarkable with no scleral icterus or jaundice and a normal cardiovascular and respiratory exam. Laboratory investigations showed a serum sodium of 136 mEq/L, potassium of 4.2 mEq/L, creatinine of 0.75 mg/dL, albumin of 3.0 g/dL, total bilirubin of 1.52 mg/dL, direct bilirubin of 0.94 mg/dL, AST of 85 U/L, ALT of 93 U/L, GGT of 776 U/L, ALP of 298 U/L, and LD of 1049 U/L. Calcium was significantly elevated at 15.75 mg/dL. He was admitted to the internal medicine service for further care.

The presence of hypercalcemia was surprising as it is not commonly associated with HCC. AFP was <1 and PTH was suppressed at 3; therefore, the etiology of the hypercalcemia and multifocal hepatic lesions pointed towards other metastatic cancers. An infused CT scan of his chest, abdomen and pelvis was done. It revealed no evidence of intrathoracic malignancy, but multiple hepatic lesions, and a conglomerate nodal mass near his duodenum.

A CT-guided biopsy was pursued and immunostains were positive for CD20, PAX5, CD5, BCL6, BCL2, and C-Myc consistent with diffuse large B-Cell lymphoma. HIV, Hepatitis B and Hepatitis C serologies were negative. A bone scan was performed which showed no metastatic bony lesions, and bone marrow biopsy showed normal trilineage hematopoiesis with no evidence of lymphoma. Hematology was consulted and after 5 cycles of R-CHOP, our patient has shown response to treatment with the largest of his lesions reduced in size from 7.8 cm by 5.9 cm to 2.7 cm by 2.3 cm.

Discussion: This case demonstrates the importance of having a systematic approach to the workup of a patient presenting with confusion. Given his longstanding cirrhosis, hepatocellular carcinoma with decompensation of underlying cirrhosis was initially felt to be the most likely etiology of his presentation. However, hypercalcemia, a normal AFP level and further imaging led us to an unexpected diagnosis with significant ramifications for the patient’s care and management.
A CASE OF STRONGYLOIDES STERCORALIS HYPERINFECTION TREATED WITH VETERINARY IVERMECTIN PREPARATION

Authors: Shyam Kolangara MBBS, James Ladd MD, Jeremy Gradon MD

Introduction: Strongyloides stercoralis is a parasitic infection endemic to tropical countries, usually affecting the immigrant population in the United States. The cycle of autoinfection permits its presence for decades in an immunocompetent host with only minor symptoms. However, when the host becomes immunocompromised such as in the event of a severe illness, a disseminated disease or hyperinfection syndrome can develop that often proves to be fatal.

Case Presentation: A 77 year old man from Trinidad and Tobago presented with nausea, vomiting, poor appetite, abdominal pain and altered mental status. He also noted coffee ground emesis and melena. On examination, he was cachectic with marked epigastric tenderness and decreased bowel sounds. His mental status deteriorated upon admission and he soon became unresponsive and hypotensive, which prompted a transfer to the intensive care unit for intubation, mechanical ventilation and vasopressor support. An esophagogastroduodenoscopy was done and biopsy revealed the presence of Strongyloides stercoralis. Albendazole and oral ivermectin were initiated through a nasogastric tube, but given the intestinal ileus, absorption was questionable. Therefore, measures were taken to obtain a veterinary subcutaneous ivermectin formulation and a request was submitted to the Food and Drug Administration (FDA) for compassionate use. Following the FDA approval, oral preparation was discontinued, and the subcutaneous formulation was administered along with oral albendazole. Since the patient remained intubated for 21 days, a tracheostomy and percutaneous gastrostomy tube placement were performed. He was eventually weaned from ventilator and vasopressor support, and subsequently transferred to the acute care floor.

Discussion: Critically ill patients infected with Strongyloides stercoralis can develop hyperinfection syndrome, which is dangerous and challenging to treat, especially if concomitant intestinal malabsorption is present. Since parenteral ivermectin is not approved in humans, an unorthodox approach may be needed with the use of a veterinary formulation of subcutaneous ivermectin which will require FDA approval. Thus, when dealing with a patient population at risk for Strongyloides infection, it is prudent to have different formulations of antiparasitic agents readily available and seek early FDA approval for the use of veterinary preparations.

References

Aplastic Anemia Without the Anemia

Authors: Robert DeGrazia Jr M.D., Jenny Petkova M.D., Michael Lankiewicz M.D.

Introduction: Aplastic anemia is a diagnosis that requires bone marrow biopsy and depletion of at least two cell lines. This case highlights an interesting presentation of a patient who suffered from idiopathic aplastic anemia without actually developing anemia. Further, in patients who are diagnosed with aplastic anemia, the first line treatment in those who do not have a reversible cause is stem cell transplantation. In situations, such as this one however, where a stem cell transplantation cannot be performed, cyclosporine and eltrombopag (a thrombopoietin-receptor agonist) have been shown to be effective at improving hematologic response in patients refractory to immunosuppression.

Case Presentation: A 23 year old incarcerated male presented with petechiae and mucosal bleeding. He was found to have WBC of 2.6, ANC 1000, Hb 14.3 g/dL and platelets of 3,000. His metabolic panel and PT/PTT were normal. His initial workup for common bacterial and viral infections including Parvovirus, hepatitis, HIV was negative and he underwent a bone marrow biopsy which revealed marked hypocellularity of 10% without dysplastic features. He was transferred to our institution for evaluation of possible allogeneic stem cell transplant (SCT) for aplastic anemia. His repeat marrow evaluation confirmed hypocellularity of 5% and a normal karyotype. A chest CT scan identified no mediastinal masses. He received a single transfusion of platelets prior to his transfer and during his hospitalization did not require any additional transfusions. He was evaluated for possible SCT; his sister was found to be a 10/10 identical HLA match, however she was in her first trimester of pregnancy and plans for transplantation were postponed. He was immunosuppressed with cyclosporine and eltrombopag with excellent response and normalization of his white blood cell (WBC) and platelet counts. Six months after initiation of therapy we began a slow taper of his cyclosporine. His platelet count dropped initially but later stabilized at approximately 90,000. Nine months after his initial diagnosis he remains on low dose cyclosporine 100 mg and 100 mg of eltrombopag daily.

Discussion: Patients with aplastic anemia typically present with severe anemia and low reticulocyte count however rare cases with isolated neutropenia or thrombocytopenia exist. The diagnosis requires a bone marrow biopsy with <20% cellularity and at least two of the following; a platelet count less than 20,000/microL, ANC of <500/microL and reticulocyte count less than 20,000/microL. Allogeneic stem cell transplantation is the typical treatment for young healthy patients with a matching sibling donor. In the case of a transfusion independent patient whose donor was temporarily unavailable we opted for a course of immunosuppression first. Since the patient remains in remission on his current therapy, we are planning on stem cell transplantation if he experiences a relapse.
Caution with the Contaminant: When a Dismissed Lab Result Turns Dangerous

Authors: Irma N. Hashmi, DO, Meghna C. Trivedi, MD

Introduction: Blood culture remains the gold standard for diagnosis of bacteremia. In an effort to reduce hospital length of stay, exposure to unnecessary antibiotics, and patient cost, the national target for blood contamination rate is 2-3%. However, caution must be applied when rejecting a positive blood culture as a contaminant. We present the case of a patient who presented to the emergency room (ER) multiple times for sepsis due to subacute endocarditis secondary to Group G streptococcal bacteremia following dismissal of blood culture as contaminant.

Case Presentation: An unfortunate 63-year-old female with past medical history of lumbar spinal stenosis requiring multiple back surgeries, presented to the ER from a rehabilitation (rehab) center following lethargy, decreased oral intake and fever to 39.4 Celsius. The patient had originally been admitted to rehab for persistent right hip pain and weakness. At the rehab center, blood cultures were drawn and grew Group G streptococcus in one out of four bottles. This was attributed to be a contaminant. In the ER, the patient also complained of dyspnea and computed tomography (CT) chest with contrast demonstrated bilateral pulmonary emboli. Repeat blood cultures were negative. The patient was discharged back to rehab on anticoagulation. Several weeks later the patient became confused, and outpatient workup included CT abdomen pelvis which revealed new splenic and renal infarct. The patient re-presented to the ER with tachycardia and temperature 38.1 Celsius. This time, blood cultures grew Group G streptococcus in four out of four bottles. Transesophageal echocardiogram revealed mitral valve vegetation. The patient was started on penicillin treatment. Cardiothoracic surgery was consulted who recommended for the patient to undergo CT head; the imaging study highlighted scattered bilateral hypodensities concerning for embolic lesions. MRI brain confirmed these lesions to be with hemorrhagic transformation, making cardiothoracic surgery unattainable. The patient’s hospitalization course was further complicated by cerebral vascular accident resulting in right sided weakness. The Infectious Disease team followed the patient closely, and she received six weeks of ceftriaxone treatment.

Discussion: A beta-hemolytic strain within the streptococcus family, Group G streptococcus encompasses several species including Streptococcus dysgalactiae and can be found as normal flora of the upper airway and skin. However, when the bacteria does cause infection, it leads to severe infection of joint, skin, and soft tissue. Unfortunately, our patient developed a complication most often attributed with other virulent species of streptococcus: endocarditis, with splenic, renal, and neurologic sequelae. The case serves as an important lesson that even though growth in one culture bottle is often dismissed to be a contaminant, clinical presentation demands careful reevaluation to prevent multi-organ consequences.
MARYLAND CLINICAL VIGNETTE POSTER FINALIST - ARJUN KANWAL, MD

A Case of Mid Cavitary ‘Broken Heart Syndrome’ After Assault

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Introduction: Takotsubo Cardiomyopathy (TC) was first documented in the 1990s in Japan as a reversible cardiomyopathy often observed in post-menopausal women after a recent emotional stress. Apical ballooning in the left ventricle is the classic echocardiographic observation. We present a rare case of a male patient with mid-cavitary variant of TC.

Case Presentation: A 63-year-old male presented to the emergency department after assault during which he was punched in the head and choked for 20 seconds. During the choking episode he was severely lightheaded, although never lost consciousness. Upon arrival, the patient complained of pleuritic, non-radiating chest pain. Initial troponin was elevated at 0.432 and eventually peaked at 2.05 six hours later. There were no changes on electrocardiogram and patient was admitted concerning for a Non-ST elevation myocardial infarction. Echocardiography showed ejection fraction of 40-45%. Eventually the patient underwent a coronary angiogram which demonstrated normal coronaries but mid-anterior and inferior LV akinesis and ballooning pattern consistent with the mid-cavitary variant of TC. The patient was managed with supportive care, receiving a beta-blocker and ace-inhibitor. On outpatient follow up at 1 month from index event, a repeat echocardiogram showed EF recovery (55-60%) with no wall motion abnormality.

Discussion: TC is a reversible form of cardiomyopathy seen in up to 2% of patients who have signs and symptoms of ACS. It is characterized by left ventricular systolic dysfunction and a pattern of ‘ballooning’ on left heart catheterization and/or echocardiography in absence of significant attributable coronary artery disease. In a study of 1750 patients with TC, 90% of patients were women and over 80% were apical variant. The mid-ventricular variant with apical sparing, as demonstrated in our patient, was seen in less than 15% of patients. Other uncommon types of Takotsubo are basal, focal, and global variants. This case highlights that in the setting of chest pain following physical assault, although common things such as fractured ribs, contusion, dissection need to be ruled out; it is also important to evaluate for potential cardiac etiology of the pain.
Spice putting providers in a pickle- A case report on Synthetic cannabinoid induced cardiomyopathy and rhabdomyolysis

Authors: Iman S Khan, Ji Hyun Rhee

Introduction: Spice, K2, Fake Pot, Sweet Leaf are some of the commonly used names for synthetic cannabinoids (SCs) which are chemical analogs of naturally occurring cannabinoid products. They are being increasingly used for their hallucinogenic effects, lower cost and ease of availability. They are not detected in routine urine drug assays and their detection requires specialized urine drug screens making them an attractive option for young adults. There are case reports of cardiomyopathy associated with their overdose. Herein, we discuss the case of a young male who presents with rhabdomyolysis and cardiomyopathy after an intentional overdose of Spice.

Case Presentation: A 21-year-old male with past medical history of major depressive disorder and substance abuse (heroin, marijuana, spice), was brought in by emergency medical services after being found unresponsive. His vital signs were BP 78/46 mmHg, HR 97 beats per minute, RR 20 times a minute, temperature 100F, saturating 93% on room air. On physical exam, his pupils were pinpoint and he had a Glasgow Coma Scale of 8 (Eye 2, verbal 2, motor 4) with decerebrate posturing. Labs showed pH of 6.94, bicarbonate of 12, Urine Drug Screen was positive for opiates and cannabinoids, lactate of 16.4, Creatinine of 2.3, a creatinine kinase of >22,000 and a troponin of 3.35. An EKG on admission showed sinus rhythm with short PR interval and J point elevation most noticeable in lead 1. Bedside ultrasound showed biventricular failure with severe global hypokinesis. He was intubated and started on Dobutamine and Norepinephrine drips. On day 3, his pressors could be weaned off completely and an Echo showed improved Left Ventricular function while off pressors. He was extubated on day 3. His acute kidney injury resolved with hemodynamic support with Creatinine of 0.8 on discharge. He admitted to sniffing Heroin and smoking Spice prior to admission. It was suspected he had spice induced cardiomyopathy and AKI. He was discharged home. The patient had no history of coronary artery disease, no history of alcohol use, no evidence of valvular abnormality and no signs of sepsis and the cardiomyopathy was attributed to Spice. He did not have prolonged hypotension, nephrotoxic medication use and his kidney injury was attributed to rhabdomyolysis.

Discussion: There are case reports of ST-elevation myocardial infarctions triggered by SC use; most of these have been very young patients with normal-appearing coronary arteries on angiography. SCs bind to CBR1 receptors with a higher affinity than natural cannabinoids, leading to a direct negative inotropic effect on cardiomyocytes. While most case reports of SC-induced nephropathy involve acute tubular necrosis, our patient presented with rhabdomyolysis-induced nephropathy. SCs are far more dangerous than their natural counterparts in this respect and as they require special assays for detection, health care providers should be cognizant of their use and effects.
Case Presentation: A 51-year-old Chinese man presented at a hospital due to weakness and chest pain for several days. His past medical history was insignificant, and his only medication was a Chinese herbal medication that he was taking for his heart. On presentation his vital signs were BP 101/70mmHg, HR 165 and RR 30. On physical exam he had an irregular heart rate, crackles on his chest and cold and clammy extremities. Initial workup revealed atrial fibrillation and elevated troponin. A bedside Echocardiogram showed LVEF 20%. He was taken straight to the catheterization lab where coronary angiography showed clean coronary arteries, however right heart catheterization showed that he was in cardiogenic shock. The patient was transferred to the CICU and started on pressors and inotropes. He continued to be hypotensive with unstable SVTs and was taken back to the catheterization lab the same evening for endomyocardial biopsy and placement of an intra-aortic balloon pump. The biopsy report came back 24h later reading Eosinophilic Myocarditis.

The patient was started on high-dose steroids and rheumatology and hematology consults were asked to see him. Imaging and blood work were done to find the cause of his condition but were unrevealing. He stabilized, and steroid doses were tapered, but on day 11 was found unconscious and in sustained VT. He was resuscitated but continued to have rapid ventricular and supraventricular arrhythmias and unfortunately expired.

Discussion: It was concluded that the cause of his Eosinophilic Myocarditis most likely was hypersensitivity due to the Chinese Herbal medication Kyu Shin. Kyu Shin, or its active ingredient Bufotoxin, is a toad venom that has similar characteristics as Digoxin – a medication that is known to cause eosinophilia. The case is a reminder that although many herbal medications are harmless, some may cause significant harm and even – as in our case – death.
HYPONATREMIA AND BRAIN MASS - SIADH OR SOMETHING ELSE?

Authors: Gregory Vo MD, Amteshwar Singh MD, Kellen Mulhern DO, Lauren Berninger DO

Introduction: Syndrome of inappropriate antidiuretic hormone secretion (SIADH) and cerebral salt wasting (CSW) are uncommon causes of hypotonic hyponatremia often juxtaposed given their similar lab findings and presentations (encephalopathy from a neurologic insult). Distinguishing them has paramount significance as treatment of each is diametrically opposed. To further convolute this distinction, adrenal insufficiency (AI), both primary adrenal insufficiency (PAI) and secondary adrenal insufficiency (SAI), can also present with hypotonic hyponatremia (albeit by different mechanisms); however both respond rapidly with steroid supplementation. Hypotonic hyponatremia can often be a leading presentation of PAI due to impaired mineralocorticoid production, whereas hypotonic hyponatremia secondary to SAI is rare, occurring due to the loss of cortisol’s inhibitory effect on antidiuretic hormone (ADH) secretion. We present a case of hypotonic hyponatremia, where these rare diagnoses were considered causing a diagnostic dilemma.

Case Presentation: A 68 year old man presented with altered mentation and vision changes during a weeklong Caribbean cruise. He received ceftriaxone for suspected meningitis, but ultimately required hospitalization for unremitting symptoms. On initial evaluation, he was hypertensive, febrile, and lethargic. Exam showed bitemporal hemianopsia, encephalopathy, and nuchal rigidity. Laboratory studies revealed a hypotonic hyponatremia (serum osmolality 263 mOsm/kg, sodium 125 mmol/L). Liver and kidney function were normal. CT head showed a sellar pituitary macroadenoma with MRI evidence of pituitary apoplexy. He was admitted to the ICU for close monitoring. Initially, the hyponatremia was attributed to SIADH prompting fluid restriction as he was clinically euvoletic. Urine studies also indicated SIADH as urine osmolality was inappropriately concentrated (392 mOsm/kg) with relatively high urine sodium (40 mmol/L). Overnight, hyponatremia worsened, so Tolvaptan was considered but withheld due to increasing urine output of 300cc/hr. Urine studies were now indicative of CSW as urine became more dilute (urine osmolality 141 mOsm/kg) with increasing salt loss (urine sodium 84 mmol/L). Additionally, orthostatic vital signs were positive. Consequently, normal saline was initiated to match fluid and sodium loss. A central line was placed in anticipation for hypertonic saline. Coincidently, concurrent endocrine workup revealed impending panhypopituitarism (testosterone <20g/dl, LH 0.2 mIU/L, FSH 0.3 mIU/L, TSH 0.3 mIU/L, prolactin 1.9 ng/ml, cortisol 2.3 mcg/dl) and the patient was started on hormone replacement. As the hyponatremia resolved, he underwent successful resection of the macroadenoma and recovered.

Discussion: Although the cause of the patient’s hyponatremia was likely multifactorial, based on the response to corticosteroids, it was concluded that SAI secondary to pituitary apoplexy was the primary etiology. This unique case underscored the importance of distinguishing four potentially life threatening conditions that present similarly, but each have distinct treatments that can cause considerable morbidity and mortality if not properly employed. The differential for encephalopathy should remain broad otherwise anchoring bias on infectious etiology may lead to premature closure and errors.
Elusive Diagnosis: A Rare Case of Polyarteritis Nodosa

Authors: Sridhar Arthi MD, Emmich Megan DO, Greco Barbara MD

Introduction: Polyarteritis nodosa (PAN) has become very rare since the widespread distribution of hepatitis B vaccine. The recognition and diagnosis of PAN unlike ANCA associated vasculitides is difficult, requires a high level of clinical suspicion, and either angiographic or histologic confirmation. We present a case of histologically proven PAN, which evaded diagnosis despite multiple admissions and specialist consultations in order to raise awareness of this, rare but often-fatal disorder.

Case Presentation: A 78-year-old male was admitted with fevers, night sweats for 2 months and a 10lb weight loss with post-prandial abdominal pain. He had a serum creatinine of 0.7mg/dl (eGFR 90 cc/min), a WBC count of 25,000 and platelet count greater than 1 million. He had a bone marrow biopsy done previously which had shown a hypercellular marrow. Extensive workup for infectious causes was performed including an abdominal CT scan, which showed no abnormalities and a normal renal perfusion. He was readmitted 3 weeks later complaining of severe post-prandial abdominal pain. CTA of the abdomen demonstrated mesenteric artery stenosis with vascular calcifications consistent with diffuse atherosclerosis without any aneurysms and normal renal arteries. He declined all interventions and went home on Hospice care. He presented for his next admission with continued abdominal pain, right foot weakness and now a 16lb weight loss. His creatinine was 2.3 mg/dl, WBC of 18,000 and ESR was 62. Urinalysis showed specific gravity 1.015, 1+ protein, 16 WBCs/HPF and 5 RBCs/HPF with no casts. Protein/creatinine ratio was 0.32. ANA, cryoglobulins, ANCA screen, hepatitis panel were all negative. Serum protein electrophoresis and immunofixation showed no monoclonal gammopathy but an elevated kappa/lambda light chain ratio of 2.8. An UGI series suggested a gastric ulcer. The creatinine continued to rise to 2.6 mg/dl and nephrology was consulted. A kidney biopsy was then done and showed necrotizing arteritis of interstitial arteries without necrotizing or crescentic glomerular lesions most consistent with PAN, mild ATN and interstitial fibrosis. He was initiated on IV pulse steroids and oral cyclophosphamide. However, the patient developed acute hypoxic respiratory failure due to pulmonary edema, declined intubation and passed away.

Discussion: PAN is a rare systemic vasculitis involving the medium sized vessels which is not ANCA driven and the pathophysiology is still not very clear. Acute kidney injury due to ischemia, renal infarction or intrarenal hemorrhage can occur. Renovascular hypertension may be present and arteriography may show vascular microaneurysms missed by CT angiography. Historically, PAN was often associated with hepatitis B. Most cases now are idiopathic. The French Vasculitis Study Group (FVSG) proposed the Five Factor Score (FFS) to prognosticate patients based on organ involvement. The 5-year mortality of PAN patients with FFS of 2 have a mortality of 46%. PAN should be considered in patients with multisystem disease associated with organ ischemia.
IT’S ALL ABOUT TIMING!

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Introduction: Thyroid diseases influence cardiac parameters, including cardiac contractility, myocardial oxygen consumption, cardiac output, blood pressure and systemic vascular resistance. Hypothyroidism affects about 9 to 15% of the adult female population, and it can cause both systolic and diastolic dysfunction of the heart. Hypothyroidism as a cause for cardiomyopathy is rare and is a diagnosis of exclusion.

Case Presentation: A 45 year old female with a history of hypothyroidism secondary to thyroidectomy presented to the hospital with complaints of two weeks of progressively worsening shortness of breath, associated with orthopnea, paroxysmal nocturnal dyspnea and peripheral edema. Sudden onset of non-radiating substernal chest pressure brought her to the hospital. For coexisting hypoparathyroidism, she had been taking calcium along with levothyroxine. Physical examination revealed stable vital signs, normal heart sounds, jugular venous distension, pitting edema in bilateral lower extremities up to mid-calves and crackles in the right lung base. Laboratory data revealed normal cardiac enzymes, a TSH of 70 uU/ml and normal free T4 of 1.07 ng/dl. A transthoracic echocardiogram revealed a reduced ejection fraction of 10%. Subsequent cardiac catheterization showed normal coronary arteries. She was educated to separate levothyroxine from calcium by 4 hours and started on guideline directed medical therapy with metoprolol succinate, captopril, spironolactone and furosemide. A cardiac MR revealed non-ischemic cardiomyopathy with no active inflammatory signs of myocarditis or infiltrative disease and, therefore, endomyocardial biopsy was not performed. Upon clinical improvement, she was discharged with a life vest. A follow up echocardiogram three months later showed remarkable improvement in the left ventricular function with an ejection fraction of 45% with near normalization of thyroid function tests.

Discussion: Thyroid hormones affect cardiac function by both genomic and non-genomic effects. The decreased contractility associated with hypothyroidism is due to reduced expression of sarcoplasmic reticulum Ca2+ ATPase and increased expression of its inhibitor, phospholamban. With normalization of thyroid function, an improvement in cardiovascular functional measures is expected. In the light of this case, appropriate timing of levothyroxine and consideration for interaction with other medications cannot be emphasized enough.
An Unfortunate Turn of Events: A Rare Case of Splenic Hematoma Following Colonoscopy for Post-Polypectomy Bleeding

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Introduction: Post-polypectomy bleeding has been reported to occur in 0.3 to 6.1% of colonoscopic polypectomies. Splenic injury after colonoscopy is a rare complication with a described incidence of less than 0.017%. We report a patient who developed hypovolemic shock due to splenic hematoma following a colonoscopy performed for post-polypectomy bleeding.

Case Presentation: The patient is a 66-year-old male with a past medical history significant for coronary artery disease, hypertension, type 2 diabetes mellitus and head/neck cancer (in remission post chemoradiation) who was admitted to the hospital with post-polypectomy bleed. He underwent an initial colonoscopy in July 2018 for history of adenomatous polyps. The colonoscopy revealed multiple non-bleeding polyps (about 15) throughout the large bowel and 8 polyps were removed with endoscopic mucosal resection (EMR). A follow up colonoscopy was performed for resection of the remaining polyps in October 2018. He presented to the hospital one-week post second colonoscopy with acute onset of hematochezia. On presentation, he was afebrile, with HR 64 and BP 108/62. Laboratory evaluation was notable for anemia with a hemoglobin of 7.1 g/dl (from a baseline of 11 to 12 g/dl) concerning for post-polypectomy bleeding. Colonoscopy repeated after admission (performed without difficulty) showed no active bleeding; a clean based ulcer was noted in the transverse colon at the site of EMR which was the likely source of bleeding. Hospital course was complicated by severe left upper quadrant pain and shock with BP dropping to 70s/40s requiring ICU admission one day after the colonoscopy. He was found to be coagulopathic with an INR of 4.9 requiring vitamin K and FFP (INR was 1.1 at time of the colonoscopy). Hemoglobin dropped to 5.3 g/dl and he received a total of 10 units of pRBC transfusions. CTA abdomen/pelvis showed a splenic hematoma measuring 12.3 x 10 x 17 cm with extravasation of the contrast during venous phase. Splenectomy was deferred per surgery recommendations and coil embolization of the proximal splenic artery was successfully performed. He improved clinically after splenic artery embolization and was discharged home with close follow-up.

Discussion: Splenic injury is a potentially catastrophic complication of colonoscopy with a reported mortality of about 5%. Several mechanisms of splenic injury associated with colonoscopy have been described which include difficult procedure, traction on the splenocolic ligament, external abdominal pressure during colonoscopy, perforation of the splenic flexure and direct trauma to the spleen, and traction of adhesions between the spleen and colon. Patient-related risk factors such as abdominal surgeries leading to adhesions, splenomegaly and pancreatitis increase the likelihood of splenic injury during the procedure. Although rare, an increasing number of cases have been reported in the last decade and further studies are required to assess the incidence and risk factors for splenic injury after colonoscopy.
MASSACHUSETTS CLINICAL VIGNETTE POSTER FINALIST - ALI ALKHAYAT, MD

Medical Therapy Refractory Salt–Sensitive Hypertension: Liddle Syndrome, Late Presentation

Authors: Ali Alkhayat, MD; Uliviyya Gasimova, MD, PhD; Dina A. Ibrahim, MD; Salaheldin Elhamamsy, MD, FACP

Introduction: Liddle’s syndrome, a rare hereditary disease, is characterized by early-onset of salt-sensitive hypertension, hypokalemia, metabolic alkalosis, low plasma renin activity and hypo-aldosteronism, that begins unusually early in life, often in childhood caused by disproportionate salt and water reabsorption at the distal nephron. We present a case of liddle syndrome presented in the outpatient setting in his sixth decade of life, with no family history of the disease.

Case Presentation: A 53-year-old African American male, with medical history of diabetes mellitus presented with a diagnosis of hypertension. At the initial visit, his blood pressure was 178/106 mmHg without significant asymmetry or postural variation. His medications included Amlodipine 10 mg. The initial lab workup showed creatinine level was 1.79 mg/dl, blood urea nitrogen 27 mg/dl, serum sodium 141 meq/L, serum potassium 3.5 meq/L, carbon dioxide 28 meq/L, and proteinuria. However, despite receiving various combinations of angiotensin-converting enzymes inhibitors, thiazide diuretics, Beta blockers all at high doses, along with sodium-restricted diet failed to control his blood pressure. Follow up visits revealed persistent hypokalemia. Additional workup showed serum Aldosterone 4.5 ng/dl (upright position), plasma renin activity 3.7 ng/ml/hr (upright position), serum sodium 142 meq/L, serum potassium 3.2 meq/L, and carbon dioxide 33 meq/L. Serum cortisol, serum and urinary metanephrine, serum protein electrophoresis, complement levels, and P-ANCA and C-ANCA were all within normal levels. Doppler Ultrasound did not show renovascular disease. Response to Amiloride without any recurrence of hypertension and hypokalemia confirmed the diagnosis of Liddle’s syndrome.

Discussion: In a patient with hypertension, hypokalemic alkalosis, and hyporeninemic hypo-aldosteronism, the possibility of Liddle’s syndrome should be considered. Liddle’s syndrome is a rare autosomal dominant condition characterized by increase in sodium reabsorption from the collecting tubule and secretion of potassium in the majority of the cases. Genetic studies have revealed that mutations affecting the cytosolic tail of the β subunit of the epithelial sodium channel (ENaC) could lead to this disorder. These mutations apparently cause constitutive activation of the epithelial sodium channel of the luminal membrane of the collecting tubule and result in excessive absorption of sodium, leading to volume expansion. This in turn causes hypertension, leading to inhibition of renin and aldosterone secretion. Lack of down-regulation of the epithelial sodium channels despite persistent volume expansion is the basis behind the pathogenesis of this syndrome. Patients mostly present at a young age, although occasionally cases may not be detected until adulthood. However, presentation in the 6th decade of life or later has been reported very rarely. It is important to screen for this condition in patients with hypertension, hypokalemia and metabolic alkalosis, as the treatment of Liddle’s syndrome differs from other forms of essential or secondary hypertension. Potassium-sparing diuretics such as Amiloride and Triamterene, which directly close sodium channels, are effective in Liddle’s syndrome.
I have a fever and the only cure is the right diagnosis

Authors: Jose R. Aponte-Pieras, MD; Aleena Paul, MD; Meghna Trivedi, MD

Introduction: We present a patient with bioprosthetic aortic valve, history of recurrent dental restorative work for poor dentition initially presenting with cerebellar stroke found to have Streptococcus mutans prosthetic valve endocarditis.

Case Presentation: A 64-year-old male with a history of aortic valve stenosis status post bioprosthetic valve replacement nine months prior, paroxysmal atrial fibrillation on warfarin, and extremely poor oral hygiene presented to the hospital with acute onset of vertigo and left sided weakness. An acute cerebellar infarction was diagnosed despite a therapeutic INR on admission. He subsequently developed rapid atrial flutter refractory to chemical cardioversion. TEE cardioversion was successful and demonstrated normal appearing valves. Detailed history revealed fatigue, low grade fevers, night sweats, and weight loss for five weeks preceding admission. On hospital day three, he had a fever of 38.6 degrees Celsius. Chest X-ray and urinalysis were unremarkable; blood cultures were not drawn until the day of discharge. Patient was readmitted two days later as four sets of blood cultures grew Streptococcus mutans. Physical examination on readmission revealed multiple dental caries, missing teeth, a two out of six systolic murmur best heard near the right sternal border (not documented in physical exams on the initial admission), mild left sided ataxia and dysmetria. Patient also reported his last visit for restorative dental work to have been four months prior. Transesophageal echocardiogram on readmission revealed a vegetation on the bioprosthetic aortic valve. CT chest ruled out an aortic root abscess. Infectious disease consultation recommended treatment with ceftriaxone given high penicillin susceptibility. Cardiothoracic and oral surgery recommended no surgical intervention. A repeat TEE after six weeks of antibiotic therapy confirmed resolution of endocarditis.

Discussion: Streptococci are amongst the most common pathogens causing prosthetic valve endocarditis (PVE) less than a year postoperatively. The most common symptom in infectious endocarditis is fever (up to 90% of patients), and 85% will have a cardiac murmur. 20-40% of PVE cases are complicated by central nervous system manifestations of septic emboli. Blood cultures are positive in 90% or more patients with PVE, while TEE has 86-92% sensitivity. Streptococcus Mutans, a member of the viridans family of alpha hemolytic streptococci, is a common pathogen in tooth decay causing up to 17% of prosthetic valve endocarditis cases. Invasive dental procedures can contribute to the risk of infectious endocarditis. In patients with prosthetic valves and cardiac risk factors, the risk of endocarditis following a dental procedure can be almost five times higher than native valves. This case demonstrates the importance of clinical suspicion needing to align with objective clinical data to ensure there is no misdiagnosis. The patient’s subacute constitutional symptoms, prior valve replacement, and associated fever should have led to consideration of endocarditis as the etiology of the initial acute stroke.

References

“Urine” for a Shocking Diagnosis: Pigment Nephropathy as an Uncommon Complication of Sulfadiazine in Treatment for CNS Toxoplasmosis

Authors: Michael Devine DO, Meghna Trivedi MD, Department of Medicine, University of Massachusetts Medical School, Worcester, MA

Introduction: Pigment Nephropathy due to sulfadiazine therapy is a rare complication of treatment for CNS *Toxoplasma gondii*. The broad constellation of presenting signs and symptoms combined with a relative paucity of cases can make accurate and timely diagnosis challenging.

Case Presentation: A 35-year-old HIV positive gentleman presented to the hospital with acute bilateral flank pain associated with dysuria and “sandy pink” urine. He was diagnosed and treated for central nervous system (CNS) *Toxoplasma gondii* with sulfadiazine and pyrimethamine approximately one month prior to this hospitalization. Vitals signs were stable. Physical exam revealed right abdominal, suprapubic, and flank tenderness to palpation. Diagnostic tests were notable for a creatinine of 1.9 mg/dL (baseline creatinine of 1.2), and urinalysis showed cloudy urine with trace leukocyte esterase, 1+ proteinuria, and 3+ blood. Urine culture was sent on admission. A computed tomography (CT) scan of the abdomen and pelvis with contrast showed findings suggestive of bilateral pyelonephritis without evidence of abscess, perinephric collection, or hydronephrosis. He was admitted to the medicine service for pyelonephritis and acute kidney injury. Intravenous Piperacillin-Tazobactam was started. Renal function continued to worsen despite fluid resuscitation and antibiotics. A renal ultrasound demonstrated a non-obstructing stone at the midportion of the left kidney with an equivocal correlate on CT scan of the abdomen. Antibiotics were discontinued after urine cultures showed no growth. Urine sediment was notable for “shock of wheat crystals”. Analysis of stones from the strained urine yielded N4-Acetyl-Sulfadiazine crystals. Sulfadiazine was discontinued and the patient was started on a regimen of clindamycin and pyrimethamine for CNS toxoplasmosis. The patient was given aggressive intravenous fluids and furosemide and his creatinine improved to 1.2 at the time of discharge.

Discussion: Obstructive acute renal failure associated with sulfadiazine has been reported since the 1940s. Sulfadiazine is acetylated in the liver and excreted in the urine. Low aqueous solubility leads to precipitation in the urine and subsequent mechanical abrasion, chemical irritation, and tubular inflammation. One retrospective study reviewed 35 Acquired Immunodeficiency Syndrome (AIDS) patients from 1987 to 1995 and found an incidence of sulfadiazine associated renal impairment of 1.9%-7.5% with a median onset after roughly 3 weeks of treatment (1). Principles of therapy include rehydration with a goal urine output of 1.5 Liters per day along with urine alkalinization with a goal pH of at least 7.15. In this particular cohort, nearly 90% of patients showed substantial or complete renal recovery over a median of 6 days after initiating treatment. Sulfadiazine can either be continued at a lower dose or substituted with clindamycin and pyrimethamine. This case serves as an important reminder to recognize medications as a potential etiology of acute kidney injury and abdominal pain in HIV positive patients receiving treatment for CNS Toxoplasmosis infection.

References

Brugada Pattern: Transformation During Fever

Authors: Ian Andrew Downs, MD, Anshul Srivastava, MD

Introduction: Brugada syndrome is a rare inherited disorder that is associated with an increased risk of ventricular arrhythmias (VA) and sudden cardiac death (SCD). It is defined by two specific ECG morphologies that require prompt diagnosis. Brugada pattern occurs when the ECG criteria is present without clear symptoms. Here we present a case of Brugada transformation from one pattern to the other during fever.

Case Presentation: A 35 year-old man with an ECG history of Type 2 Brugada pattern presented to the emergency department with sub-acute knee pain and malaise. He was found to be febrile, and knee aspiration diagnosed septic arthritis. ECG showed a normal rate, with transformation of his prior Type 2 Brugada pattern to a Type 1 pattern. He denied any history of syncope, palpitations, or exercise intolerance but endorsed early death in his father at 30. He defervesced with the start of scheduled anti-pyretics and antibiotics but remained in a Type 1 Brugada pattern.

He was admitted to the hospital with cardiac monitoring. A transthoracic echocardiogram showed a structurally normal heart. He underwent an uncomplicated arthroscopic washout while avoiding medications known to trigger deterioration of Brugada pattern into VA. The patient was discharged with a course of antibiotics, an event monitor, and close follow up for consideration of implantable cardioverter-defibrillator (ICD) placement.

Discussion: The Brugada syndrome and pattern are defined by characteristic ECG findings. The Brugada pattern is differentiated from Brugada syndrome by lack of clear syncopal symptoms or VA. In type 1, the ST segment shows an upward convexity, or “coved” appearance, while in type 2 it shows a downward convexity, or “saddleback” appearance. The type 1 pattern carries higher risk for transformation to VA.

The pattern is inherited in autosomal dominant fashion due to mutations in cardiac myocyte channels. Fever also affects cardiac repolarization and is a trigger for VA and SCD in Brugada. In a retrospective review, fever preceded SCD in 4 of 22 patients (18%) with Brugada syndrome. In a case-control study, the prevalence of type 1 Brugada pattern was significantly higher in febrile ED patients than afebrile patients (2% vs 0.1%).

No prospective data for the management of fever-induced Brugada pattern changes exists, but aggressive management of the fever is the mainstay of treatment. Avoidance of drugs known to provoke VA is also central. This was particularly relevant due to our patient’s need for emergent surgery, and because common anesthetics, including propofol, can trigger VA.

Long term, ICD placement is the mainstay of treatment to avoid SCD. The early death of the patient’s father was concerning for SCD, but after risk-benefit discussion with the patient, the decision was made to place an event monitor for further risk stratification before ICD placement.

References

A Rare Case of Concurrent Ankylosing Spondylitis and Mixed Connective Tissue Disease Successfully Treated with Adalimumab

Introduction: The concurrence of spondyloarthopathies like ankylosing spondylitis (AS) and connective tissue diseases like mixed connective tissue disease (MCTD) has been rarely described (with 3 reported cases in the literature). Adalimumab has demonstrated good efficacy and safety profile for the treatment of AS but has not been studied in MCTD. We report a patient diagnosed with both AS and MCTD who was successfully treated with adalimumab after failure of therapy with NSAIDs and non-biologic regimens.

Case Presentation: The patient is a 24-year-old Japanese male who initially presented to the rheumatology clinic in 2014 with a 6-month history of pain and morning stiffness involving the lower back, pain and swelling of hands/feet, diffuse myalgias with lower extremity proximal muscle weakness, and bluish discoloration of fingers and toes on exposure to cold. Work-up back then revealed positive ANA with a high titer of 1:1280 and speckled pattern on immunofluorescence; negative anti-dsDNA; high positive anti-U1 RNP [7.9 AI, normal < 1 antibody index (AI)]; positive anti-Sm (1.5 AI), anti-Sm/RNP (> 8.00 AI), and anti-chromatin antibody (>8.0 AI); positive HLA-B27 and elevated CK (427 U/L, normal range 44-196 U/L). He tested negative for anti-Jo1, anti-Mi2, anti-Scl 70, anti-Ro and anti-La antibodies. An MRI of sacroiliac joints showed bilateral sacroiliitis. At the age of 20, he was diagnosed with AS based on ASAS criteria of inflammatory back pain, positive HLA-B27 and bilateral sacroiliitis on MRI. He was diagnosed with MCTD based on both Alarcon-Segovia’s and Kahn’s criteria of high titer anti-RNP, Raynaud’s phenomenon, myositis, synovitis, and swollen hands/fingers. On pulmonary function testing, he had borderline restrictive lung disease with mildly reduced DLCO. He had a good response to NSAIDs, however, his symptoms recurred. He was trialed on methotrexate, hydroxychloroquine, and prednisone without significant response. Subsequently, he was started on adalimumab in 2015 with marked improvement in back pain and range of motion, resolution of muscle weakness, synovitis and finger swelling. CK normalized 5 months after treatment with adalimumab. At the follow-up visit in 2018, his symptoms were well controlled with adalimumab monotherapy without any significant adverse effects. However, he continued to have Raynaud’s, worse in the winter and was treated with nifedipine.

Discussion: AS is an axial spondyloarthritis typically affecting the spine, which often presents with back pain. On the other hand, MCTD is a rare connective tissue disorder characterized by overlapping features of systemic lupus erythematosus, systemic sclerosis, and polymyositis with a high titer of anti-U1 ribonucleoprotein antibodies. This case demonstrates successful treatment of refractory AS and MCTD with adalimumab. Treatment with TNF-alpha inhibitors like etanercept and infliximab has been described previously in case reports to control refractory polyarthritis in individuals with MCTD. Further studies are required to assess the efficacy of adalimumab in patients with refractory MCTD.

References


IgM Multiple Myeloma Disguised as Waldenstrom's Macroglobulinemia

Authors: Lauren Mendelson, D.O.; Nattamol Horsiriluck, M.D.; Venu Bathini, M.D.

Introduction: Immunoglobulin (Ig)M Multiple Myeloma (MM) is a rare disease entity that can present with many symptoms and diagnostic features that overlap with Waldenstrom's Macroglobulinemia (WM). Despite a similar presentation, the management, treatment, and prognosis of the disease processes differ entirely. Achieving the correct diagnosis is a difficult but essential task.

Case Presentation: We present an 84-year-old female with remote history of stage IIc ovarian carcinoma and recent diagnosis of retinal hemorrhage who presented with subacute onset of nose bleeding, gingival bleeding, and dyspnea on exertion. She was found to have lab values consistent with IgM hyperviscosity syndrome with serum viscosity: 4.7 relative to water (reference range 1.5-1.9), IgM: 7,000mg/dL, and total protein: 7g/dL. Bone marrow biopsy revealed a B-cell lymphoproliferative disorder with 38% cluster of differentiation (CD)56 negative plasma cell differentiation consistent with WM. The patient was treated for WM with emergent plasmapheresis and Bendamustine. Cytogenetics then revealed a positive MAF-IgH (14:16) translocation and a negative MYD88 mutation confirming the diagnosis of IgM MM rather than WM. Treatment was switched to bortezomib, cyclophosphamide, and dexamethasone. After induction chemotherapy she declined bone marrow transplant and had repeat bone marrow biopsy which showed 3% plasma cells and negative MAF-IgH (14:16) translocation. She was started on maintenance therapy with lenalidomide which she will continue until disease progression or toxicity development.

Discussion: This case illustrates the importance of distinguishing between WM and IgM MM. Clinical features and bone marrow cell morphology are used as the first line differentiation; however, due to the overlapping features, cytogenetics should be used to further delineate the diseases. It is crucial to make the correct diagnosis as WM is treated with rituximab to target B-lymphocytes, whereas IgM MM is treated with induction chemotherapy followed by autologous stem cell transplantation. Differentiation between WM and IgM MM is critical as treatment and prognosis differ drastically.

References

MASSACHUSETTS CLINICAL VIGNETTE POSTER FINALIST - MASOOD PASHA SYED, MD

Break-up Before Birth

Authors: Masood Pasha Syed, MD (Associate), Adhirath Doshi, MD, Srinivas Nadadur, MD, Mohamed Hassan, MD, Robert Harizi, MD, Department of Medicine, Saint Vincent Hospital, Worcester, MA

Introduction: Isolated left ventricular noncompaction (LVNC) can be sporadic or familial and is characterized by prominent trabeculae and deep intertrabecular recesses. It has been found in 0.014-1.3 percent of patients undergoing echocardiography, and it is an uncommon cause of heart failure and sudden cardiac death.

Case Presentation: A 59-year-old male with a history of COPD presented with dyspnea and leg swelling over 3-4 weeks. His baseline COPD-related exertional dyspnea progressed to symptoms at rest along with new orthopnea. He reported no chest pain, palpitations, syncope, fever, chills, wheezing or a productive cough. His mother and brother died at ages 54 and 44, respectively, after developing heart failure. The patient smokes 4-5 cigarettes a day, with a 40 pack-year history. Examination on admission revealed stable vitals, elevated JVP, normal S1, S2 regular heart rate, diminished air entry in bilateral bases with occasional wheezing and bibasilar rales. His lower extremities revealed bilateral 1+ non-tender pitting edema with clubbing.

The EKG showed sinus tachycardia with a right axis deviation with evidence of left ventricular hypertrophy and right atrial enlargement. There were no ischemic changes. CXR was normal. An echocardiogram showed global hypokinesia with a left ventricular ejection fraction of 35%-40% with grade I diastolic dysfunction and probable noncompaction of the left ventricle. Right ventricular function and right ventricular systolic pressure were normal. CAD risk factor screening was significant for smoking and a provisional diagnosis of left ventricular non-compaction (LVNC) was made.

He was started on furosemide, lisinopril and carvedilol. A right heart catheterization revealed non-ischemic cardiomyopathy secondary to LVNC with a mildly elevated pulmonary wedge pressure. Cardiac MRI confirmed increased trabeculation in both ventricular apices with impaired biventricular systolic function, confirming non-compaction cardiomyopathy.

Discussion: LVNC is thought to be due to an intrauterine arrest of compaction of the loose interwoven meshwork present in fetal myocardial primordium. Alternatively, the pronounced hypertrabeculation may be due to altered regulation in cell proliferation, differentiation, and maturation during ventricular wall formation. Clinically, it may have variable manifestations of heart failure, chest pain, thromboembolic events, and atrial and ventricular arrhythmias, including a risk of sudden cardiac arrest (SCA). Our patient presented with new onset heart failure with decreased systolic function, but clean coronaries on cardiac catheterization.

The significant family history pointed us towards a cardiomyopathy workup. A diagnosis of non-ischemic cardiomyopathy due to LVNC posed our patient to the risk of arrhythmias, thromboembolism, and SCA. He was advised to undergo Holter monitoring following discharge to screen for arrhythmias, discuss the role of an implantable cardioverter-defibrillator, anticoagulation given reduced ejection fraction and genetic counselling.
MEXICO - CLINICAL VIGNETTE POSTER FINALIST - DANIEL REBOLLEDO

A man with retroperitoneal bleeding and coagulopathy: an unusual presentation in amyloidosis spectrum

Authors: Daniel Rebollo García, Luis Servando Jimenez-Reyes, Gabriela García Martinez, Jesus Duarte Mote. MD, Ruth Gutierrez Serdán

Introduction: Wherever chronic inflammatory states can trigger systemic amyloid material deposition in several organs. Hematological manifestations of this entity are unusual, thus, it’s necessary a structured approach in to findings. In Mexico, Hernández-Reyes et al. For 30 years ago, report a serie of 23 patients, multiple myeloma was the most frequent cause, interesting it’s prevalence is 14 times less with respect to caucasian series, and, the mainly of topic for our population, however these case descrites without with coagulopathy presentation secondary due to amyloidosis.

Case Presentation: A 48-year-old man, country person, with a medical record of plastic surgery due to correction to harelip at age 9 years old and exposure to pesticides and herbicides at least 12 years previously, arrive to emergency room due to syncope and abdominal pain of a suddenly evolution. He presented hypovolemic shock; findings of surgical laparotomy was a retroperitoneal haematoma of 1,500 cc without secondary findings, thus he required management in the ICU. Afterwards, an abdominal CT scan report a waste retroperitoneal haematoma without secondary cause that was considered "spontaneous" and another relevante finding was hepatomegaly and splenomegaly. Quantitative and functional hematological blood test were unremarkable, however, a widening of coagulation time in prothrombine time, INR and tromboplastin time were the most important finding, that were corrected with blood plasma and cryoprecipitates with a short duration in the effect and continued in persistent widening and retroperitoneal bleeding, it was necessary to made an approach to found disturbs in coagulation factors, that result in a coagulation factor X activity less than 5 percent (normal value 70- 120%).

Discussion: Approach in this case included most frequent etiologies of bleeding in Mexico (oral anticoagulants, liver and kidney diseases); due to lack of resources in our hospital, we use epidemiology studies to found deficiencies and disorders in coagulation factors that affect the common pathway of the coagulation cascade how in this case. Low activity of factor X based on background and context of this case on lympho and myeloproliferative processes, multiple myeloma and amyloidosis. Results of specific haematological test were normal peripheral blood smear, five percent of plasmatic cells in the bone marrow aspirate, serum light proteins (Kappa) in 230 mg (3.3-19.4), serum protein M in 1.62 gr (<1.5 gr), Congo red stain positive in periumbilical fat and urinary monoclonal protein electrophoresis in 328 mg (<500), match with the spectrum of monoclonal gammopathies. Final diagnosis was secondary light-chain amyloidosis due to monoclonal gammopathy of undetermined significance (MGUS).

This case ilustrated importance of semiology and a good anamnesis that remained how "gold standard" approaches of atypical presentation of common diseases, however age of debut in this case is extremely rare, because the peak of MGUS is 70 years old in average.

References

Catastrophic antiphospholipid syndrome complicated by intracranial hemorrhage

Authors: Anju Adhikari, MD, Shrinjaya Thapa, MD, Dilip Khanal, MD

Introduction: Catastrophic Antiphospholipid Syndrome (CAPS) is a rare variant of Antiphospholipid syndrome (APS). It represents less than 1% of all antiphospholipid syndrome (APS) and has mortality rate of 50%. It is characterized by diffuse thrombotic microangiopathy in the presence of antiphospholipid antibodies. As opposed to large vessel thrombosis that is typically seen in APS, pathogenesis involves diffuse small vessel thrombosis. Patients typically present with rapidly progressing multiorgan failure.

Case Presentation: 38-year-old female with the past medical history of APS, multiple spontaneous abortions and recent lower extremity deep vein thrombosis on Enoxaparin presented with nausea, vomiting and fever for ten days. She further endorsed fatigue, weight loss and decreased appetite following a recent trip to Albania. Her vitals showed a fever 101°F, tachycardia, and a borderline low BP. Rest of physical exam unremarkable except for lethargy and generalized weakness. Labs were significant for acute thrombocytopenia, chronic anemia, acute kidney injury, and elevated transaminases and troponins. Given her symptoms, CT abdomen/pelvis was done which was unremarkable. A broad infectious workup was done which was non-revealing as well. On day 3, patient developed confusion, left arm weakness and seizures, and was transferred to the medical ICU. CT head revealed right parenchymal and subarachnoid hemorrhages. Hematology testing showed elevated PT, aPTT and D-dimer. Disseminated intravascular coagulation and thrombotic thrombocytopenic purpura were ruled out the help of with normal fibrinogen, negative hemolytic panel, and absence of schistocytes on a peripheral blood smear. Anticardiolipin and Beta-2-glycoproteins antibodies were noted to be elevated. With a suspicion of CAPS, high dose IV Steroids and IV Ig were started. Plasma exchange was avoided given the associated risk of worsening of Intracranial bleed. Patient showed clinical improvement after 3 days of treatment and was successfully transitioned to Rituximab.

Discussion: CAPS is believed to mediated by immune activation leading to SIRS and widespread small vessels thrombosis. Common manifestations include fever, fatigue, acute renal failure, pulmonary edema, acute respiratory distress syndrome, stroke and transient ischemic attack (TIA). Cerebral involvement is fairly common and seen in around 62% of patients with CAPS. A definite diagnosis of CAPS requires biopsy proven small vessel thrombosis in addition to positive APL antibodies and multi-system involvement. However, treatment should not be delayed where there is high clinical suspicion as early treatment can be life-saving.

Although thrombosis is the major manifestation, bleeding can occur in 10% of patients. Bleeding is seen in patients with anticoagulation use, capillaritis, thrombocytopenia or anti-prothrombin antibodies. In such cases, treatment benefit with anticoagulation should be weighed against the risk of bleeding. High dose steroids and anticoagulation is the first-line treatment for CAPS, while IVIG infusion and plasma exchange are reserved for severe CAPS. Despite treatment, CAPS has shown to have high mortality approaching 50%.

References

Impending Rupture of an Infected Abdominal Aortic Aneurysm - A Rare Sequelae of Salmonella Bacteremia

Authors: Ali Ahmad, M.D., Halina Kusz, M.D. F.A.C.P.

Introduction: Infectious aortitis is a very rare and life threatening condition caused by microorganisms in the aortic wall which can precipitate aortic aneurysm and rupture. Infected aortic aneurysms only account for about 0.7-2.6% of all cases of aortic aneurysms. We present a rare case of an abdominal aortic aneurysm that was complicated by Salmonella bacteremia, leading to infectious aortitis and impending rupture.

Case Presentation: A 56 year old Caucasian male with a prior history of chronic obstructive pulmonary disease (COPD) presented to the hospital with shortness of breath, productive cough, fever and chills. He was found to be in acute exacerbation of COPD secondary to right basilar pneumonia, and treatment was initiated. As part of the initial workup, a CT scan of the chest and abdomen was ordered, which revealed an incidental finding of an infrarenal abdominal aortic aneurysm, measuring 5.1 x 5.1cm x 10.4cm. Blood cultures ordered on admission returned positive for Salmonella enteritidis, which was sensitive to ceftriaxone. Appropriate antibiotic treatment was initiated. However, the patient subsequently developed abdominal discomfort and new fever spikes. Repeat lab findings showed an increased leukocyte count. CT angiogram was repeated and demonstrated a significantly increased size of the aneurysm to 7.6 x 9.1 x 13.3cm, concerning for impending rupture and underlying aortitis. Nuclear white blood cell scan was performed and revealed abnormal increased uptake involving the mid and distal abdominal aorta, suggestive of infection. Vascular consultation was obtained and percutaneous endovascular aortic repair was successfully performed. He tolerated the surgery well and observed improvement in symptoms, following which he was discharged.

Discussion: Infectious aortitis has a significant mortality rate ranging from 11 to 36% in patients who are treated with both antibiotics and surgery, compared to upwards of 90% in patients treated without surgical intervention. Risk factors for infectious aortitis include age above fifty years, male gender, diabetes mellitus, hypertension, coronary artery disease, and atherosclerosis of the aorta. Symptoms may include fevers, chills, back and abdominal pain. It is associated with rapid growth and a high risk of rupture. This case demonstrates that patients with an existing abdominal aortic aneurysm who develop Salmonella bacteremia are particularly prone to infectious aortitis and rapid expansion of the aneurysm. If not diagnosed in time, it can lead to aneurysmal rupture, with life-threatening consequences. It is therefore imperative to initiate early antibiotic treatment with prompt surgical intervention.
Cerebral Beads Embolism after Transarterial Chemoembolization. A Serious Side Effect of a Relatively Safe Procedure

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Introduction: Transarterial chemoembolization (TACE) is a procedure commonly used for treatment of unresectable hepatocellular carcinoma (HCC). It involves the injection of lipiodol with or without a chemotherapeutic agent into the tumor feeding artery to disturb its blood supply and ultimately regress the tumor. Although TACE is generally considered a relatively safe procedure, serious side effects can occur. We present a case of cerebral beads embolism (CBE); an extremely rare side effect following TACE.

Case Presentation: A 64-year old man with biopsy-proven, poorly differentiated, locally advanced HCC, presented to the hospital for an elective TACE procedure. Medical history was significant for liver cirrhosis and previously treated hepatitis C and HIV with undetectable viral loads and CD4 cell count 449.

Following the procedure, his mental status became altered and he exhibited left-sided hemiparesis. A CT scan of the head without contrast was inconclusive. A follow-up MRI of the brain showed multiple punctate restricted diffusion lesions throughout both cerebral hemispheres, the brain-stem, and both cerebellar hemispheres; surrounded by vasogenic edema. These findings were consistent with acute embolic infarcts. Continuous cardiac telemetry did not record any evidence of atrial fibrillation. A two-dimensional trans-thoracic echocardiography and trans-esophageal echocardiography did not show any evidence of thrombosis in the left ventricle or left atrial appendage. He was diagnosed with CBE. Notably, TACE was conducted by mixing 150 mg of doxorubicin with LC Beads® (100-300 micro-millimeter in size made by BTG Interventional Medicine, UK). LC Bead® is a deformable microsphere that consists of a biocompatible, sulphonate-modified, N-Fil hydrogel. Over his hospitalization, the patient gradually regained strength in the left side of the body and was transferred to sub-acute rehab. Unfortunately, he passed away two months later due to end stage liver cirrhosis complicated by gastric variceal bleeding.

Discussion: This case highlights a rare, yet catastrophic complication of TACE. Previous case reports described cerebral beads embolism used lipiodol beads. Unlike lipiodol beads, LC beads that were used in our patient dissolve quickly and don’t usually cause permanent damage as noted in our patient who recovered strength within a few days. To our knowledge, this is the first reported case of dissolvable LC beads-induced embolic stroke. The occurrence of CBE was found to be related to shunting between the tumor feeding artery and the pulmonary vein. Previous case reports described pulmonary and less commonly cardiac shunt in most the cases. In our patient, no cardiac shunt was found but pulmonary shunt wasn’t investigated as a source, yet suspected. We recommend that patients undergoing TACE should be screened for the presence of shunt be-forehand to decrease the incidence of such an unfortunate complication.

References

Non-fulminant Hepatitis A with Acute Renal Failure; A Rare Association

Introduction: Acute hepatitis A (HAV) is usually self-limiting benign disease. Extrahepatic manifestations of viral hepatitis included arthritis, vasculitis, cryoglobulinemia and acute renal failure commonly associated with Hepatitis B and C. We present a rare case of non-fulminant HAV leading to acute renal failure.

Case Presentation: 35-year old male presented with fever, jaundice, abdominal pain, diarrhea and dark urine. Physical examination was remarkable for scleral icterus, right upper quadrant tenderness without hepatomegaly. Initial workup demonstrated a creatinine 13 mg/dL, AST 1936, ALT 3558 with a total bilirubin of 13.3 mg/dl and a positive hepatitis A IgM. Treated supportively with hydration for acute kidney injury. He subsequently became oliguric without improvement of renal functions. Urinalysis demonstrated large proteinuria and blood with 11 RBCs without eosinophilia. Urine microscopy showed numerous RBCs without casts. Renal ultrasound was unremarkable. An immune-mediated renal injury was suspected prompting to obtain complement levels which were significantly reduced. ANA, CANCA, PANCA and Cryoglobulin were negative. Nephrology recommended aggressive diuresis and deferred biopsy. Creatinine slowly trended down with improved urine output without dialysis. The patient was discharged with normal liver function.

Discussion: The etiology of HAV-associated renal failure exists as an amalgamation of direct toxin, ischemic and immune-complex mediated injury. The first case of non-fulminant hepatitis A causing acute renal failure was published in 1978, 60 cases have been reported since. Given the current Michigan outbreak, internists need to be aware of extrahepatic manifestations of HAV infection including acute renal failure.
**MICHIGAN CLINICAL VIGNETTE POSTER FINALIST - KUTAYBA ALSAFADI, MD**

When Hepatitis A Goes Beyond Self-limiting: A Rare Case of DAH in The Setting of Hepatitis A Infection

Authors: Ahmad Murad M.D.; Kutayba Alsafadi M.D.; Asaad Nakhle M.D.

Introduction: Hepatitis A virus (HAV) infection is usually a self-limited illness that does not become chronic. It usually presents with non-specific symptoms, but can progress to acute liver failure (ALF) in less than 1 percent of cases. The course of ALF is variable and the mortality rate is high reaching 30% in patients who do not receive transplantation, and even higher in patients who develop acute respiratory distress syndrome (ARDS) during the course of their disease.

Case Presentation: We present a case of a 28-year-old male patient who was incarcerated for three weeks, and was later found down in his prison cell. Workup was started and showed ALF secondary to an acute HAV infection. The patient was started on conventional therapies for ALF, and was later deemed not a suitable candidate for liver transplant. His course was then complicated by a persistent fever, tachycardia, worsening respiratory function, and the development of severe ARDS. This was also complicated by worsening kidney function requiring dialysis.

After remaining stable for multiple days, his oxygen requirements started to increase, and he was noted to have frank blood in his endotracheal tube. His repeat CXR showed again worsening airspace disease, and his hemoglobin acutely dropped requiring blood transfusion. These findings raised the suspicion for diffuse alveolar hemorrhage (DAH) which was confirmed by bronchoscopy and bronchoalveolar lavage. Given this new diagnosis with worsening respiratory status the patient was started on extracorporeal membrane oxygenation (ECMO).

Extensive work-up to evaluate for other causes of diffuse alveolar hemorrhage including ANA, RF, C-ANCA, P-ANCA, C3, C4, and anti-GBM were negative. The patient was continued on the same management of his ALF, and was later weaned off ECMO and had significant improvement in his respiratory status.

Discussion: Most adults with HAV infection have symptomatic illness requiring conservative management only. ALF occurs most commonly in individuals >50 years of age, and those with other liver diseases such as hepatitis B or C. We are presenting a case of a young previously healthy male who developed ALF secondary to hepatitis A, complicated by the development of ARDS, and later DAH requiring ECMO support. Although DAH was described in the past in the setting of acute hepatitis C virus infection, it was never described in the setting of hepatitis A, making our case very unique. It is unknown whether Hepatitis A as a separate entity can lead to DAH, or it was mediated by ARDS process.
New Cancer Therapy Agents with Unexpected Adverse Events: Pembrolizumab-Induced Myocarditis

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Introduction: Immune checkpoint inhibitors (ICIs) are a new class of medications that have revolutionized cancer treatment. They significantly improve survival in a range of cancer types. The mechanism of action involves increasing antitumor immunity by blocking intrinsic down-regulators of immunity. By increasing activity of the immune system, ICIs can be associated with immune-related adverse events (irAE). The vast majority of reported irAEs involve endocrine glands, skin and the gastrointestinal tract. Cardiotoxicity is extremely rare and only limited to few cases reported in the literature. Herein, we report a rare case of Pembrolizumab-induced myocarditis.

Case Presentation: A 65-year-old female with a history of metastatic non-small cell lung cancer (NSCLC) presented with a 3-day history of increasing fatigue, dyspnea on exertion and orthopnea. She was recently started on the new immune checkpoint inhibitor agent, Pembrolizumab (Keytruda®) after developing progression of her cancer. She received a total of 4 cycles over a period of 3 months. Patient denied any cough, fevers, chest pain or edema. She did not have any history of diabetes, hypertension, hyperlipidemia, cardiovascular disease, or tobacco use. Physical examination was unremarkable except for oxygen saturation of 89% on room air. Laboratory evaluation revealed Brain Naturitic Peptide of 2827 pg/mL, Troponin 1.98 ng/mL, Alanine Aminotransferase 43 IU/L, Aspartate Aminotransferase 77 IU/L. Electrocardiogram showed left bundle branch block along with frequent premature ventricular contractions in a bigeminy pattern. Chest computed tomography angiography was negative for pulmonary embolism. Echocardiogram revealed left ventricular ejection fraction of 40% with global hypokinesis. Cardiac catheterization was negative for obstructive coronary artery disease.

A diagnosis of Pembrolizumab-induced myocarditis was made and treatment with oral Prednisone 1mg/kg/day was initiated. Patient’s symptoms improved significantly, and she was eventually discharged after 5 days with a plan to taper Prednisone down on weekly basis.

Discussion: Pembrolizumab is a programmed-death-1 (PD-1) ICI that has remarkably improved survival in patients with melanoma, NSCLC, Hodgkin’s lymphoma, urothelial carcinoma, head and neck squamous cell carcinoma and gastric cancer. It can be associated with wide range of irAE, however, cardiotoxicity is extremely rare. In a recently published review of ICI-related cardiotoxicity, 13 patients were identified in the literature. Of these cases, only 2 patients were on Pembrolizumab. Symptoms may include fatigue, chest pain, dyspnea, or pre-syncope. Most patients improve with administration of systemic corticosteroids, with a small fraction who may require advanced treatment with tumor necrosis factor-alpha inhibitors, or intravenous immunoglobulins.

In conclusion, ICIs provide an excellent addition to the world of cancer therapy, however, this is not without a cost. Adverse events from ICIs can involve multiple organ systems with some being life threatening. Physicians should be aware of these events and expect rare, unreported irAE in patients on ICIs.

References

MICHIGAN CLINICAL VIGNETTE POSTER FINALIST - RICARDO DE CASTRO, MD

A rare case of infectious endocarditis caused by Achromobacter xylosoxidans

Authors: de Castro, Ricardo Lessa., Lima, Neiberg de Alcantara., Lino, Danielli Oliveira da Costa., Melgar, Thomas A.

Introduction: Achromobacter xylosoxidans is an aerobic, motile, gram-negative rod. Infections with A. xylosoxidans have included meningitis, pneumonia, peritonitis, and urinary tract infections. However, bacteremia associated with valve endocarditis caused by A. xylosoxidans is rare and usually only seen in patients who are immunocompromised. In cases of endocarditis, surgical intervention should be considered early if the patient is clinically fit.

Case Presentation: A 19-year-old man presented with a history of approximately one month of intermittent fevers, chills, dry cough and pleuritic chest pain. He lost about 10 pounds during this period and was with progressive fatigue. He had a past medical history of interatrial and interventricular communication corrected at 11 months of age and aortic coarctation correction at age 10. On examination patient was febrile, tachycardic, with a normal blood pressure. Laboratory revealed an elevated white blood cell count and microcytic anemia. Kidney function normal. A chest x-ray showed some patches infiltrates and an enlarged cardiac silhouette. An echocardiogram was performed and revealed a filamentous structure adherent to his aortic valve and to the ventricular patch used to correct the intraventricular communication. A diagnose of endocarditis was given and he was on Gentamicin, oxacillin and ceftriaxone. Blood cultures showed the presence of Achromobacter xylosoxidans sensitive to carbapenem. The antibiotic was switched but the patient continued to have fevers after 7 days of antibiotics and a surgical procedure was scheduled. During the surgery, a vegetation was seen on his aortic valve but not in his ventricular patch. The surgery was done with no major complications. He went to intensive care using moderate doses of vasopressors and was extubated 12 hours after the procedure. On his 4th day of hospitalization, he had a spontaneous right side pneumothorax, which was drained and resolved after 3 days. He went home 10 days after his surgery and completed 28 days of carbapenem. His immediate and 30 day follow-up echocardiograms at follow-up did not show any vegetation. He was symptomatic at his six months and one-year follow-up appointments.

Discussion: Endocarditis is a dangerous and difficult to treat condition. Prolonged fevers with weight loss in patients with previous history of heart disease especially after cardiac surgeries should raise suspicion of this diagnose. Achromobacter xylosoxidans is a ubiquitous environmental gram-negative bacterium, very resistant to antibiotics. Human infections are rare with few cases of endocarditis described in literature. Mortality rates are high. Nosocomial infections predominate with a strong association between bacteremia and immunosuppression. Our case showed endocarditis caused by these uncommon bacteria in a patient with previous cardiac surgery, immune competent and with no recent hospitalizations.
Mitral Valve Endocarditis with Perforation from a Urinary Source. An Unusual Case

Authors: Figueroa Rodriguez Fernando, Davila Francisco.

Introduction: *Aerococcus Urinae* (AU) is a rare pathogen, identified as gram positive, catalase-negative coccus that grows in pairs and clusters which has been reported to mainly cause urinary tract infections (UTI), especially in elderly males. We present a case of AU endocarditis causing severe mitral insufficiency and perforation.

Case Presentation: A 55-year-old male with no significant past medical history presented with a one week history of dyspnea at rest, dysuria, urinary frequency and urgency. Upon presentation, his oxygen saturation was 90%, with tachycardia and a low grade fever. On exam he had significant bibasilar crackles as well as a holosystolic apical murmur; additionally he was found to have 2+ lower extremity edema and evidence of vascular emboli phenomena on his feet. Labs demonstrated leukocytosis with neutrophilia and evidence of acute kidney injury. The urinalysis had 3+ bacteria and >50 leukocytes. The patient was started on ceftriaxone. A 2D echocardiogram revealed a 1.5 cm mobile mass on the posterior leaflet of the mitral valve consistent with a vegetation. Preliminary blood cultures reported gram positive cocci in pairs and clusters. At day 3 AU was identified the culprit. Per Infectious Diseases recommendations, he was switched to gentamicin and penicillin G with plans for 6 week therapy. He was medically treated for acute heart failure. A subsequent trans-esophageal echocardiogram confirmed a ‘1.4cm x 2.5cm’ vegetation attached to the posterior leaflet of mitral valve. Also noted was severe mitral regurgitation and posterior leaflet perforation. One week into her hospitalization the patient acutely decompensated and underwent emergent prosthetic mitral valve replacement. He had a prolonged complicated post-operative period requiring mechanical ventilation and hemodialysis. The patient was discharged after 3 months to a subacute rehabilitation unit.

Discussion: Although rare, AU has been more frequently described lately due to new laboratory identification techniques. Known risk factors are advanced edge and urologic disease. It’s been associated with UTI, blood stream infections, phlegmon and endocarditis. Diagnosis is extremely challenging having to even rely on molecular testing at times. Treatment is usually with combination therapy (B-Lactam plus Aminoglycoside). The novelty of this cases relies, first of all, on the fact that AU Endocarditis is extremely rare, to our knowledge, there are 30 cases described since 1991. Furthermore, mortality rate is as high as 70% in the immediate period, our patient has survived for two years. There are only two other cases reported of patients less than 60 years old with this diagnosed with this entity, both of them survived the hospitalization as well, raising the question of whether age is a protective factor. It is imperative to have high clinical suspicion for this organism when considering the etiology of infective endocarditis, especially in elderly males with concomitant urinary tract symptoms.

References

A Case of Fulminant Clostridium difficile Colitis in a Health Care Worker treated with Loop Ileostomy and Intra-colonic Vancomycin Flushes.

Authors: Abhinav Garg, MD, Tooba Tariq, MD, Melissa Olken, MD

Introduction: Clostridium difficile colitis is a nosocomial infection caused by spore forming anaerobic bacteria Clostridium difficile which can range from mild diarrhea to toxic megacolon with perforation. Health care workers (HCWs) are at higher risk for Clostridium Difficile colitis and might also serve as asymptomatic carriers and reservoirs. Here, we present a case of fulminant Clostridium difficile colitis with toxic megacolon in a health care worker with no recent antibiotic exposure which was successfully treated with diverting loop ileostomy and intra-colonic vancomycin flushes.

Case Presentation: A 55-year-old, emergency department nurse presented with 10-day history of diffuse, 8/10 abdominal pain and profuse, watery diarrhea. She had no prior antibiotic use or sick contacts. Pertinent physical exam finding included tachycardia, moderate, diffuse abdominal tenderness with hyperactive bowel sounds. Lab work was significant for leukocytosis (WBC count of 54,000) and CT abdomen/pelvis with contrast revealed pan-colitis with suspicion for toxic megacolon. She was started on ciprofloxacin, metronidazole and PO vancomycin. General surgery consulted and patient transferred to surgery ICU for closer monitoring and serial abdominal examinations. Antibiotic regimen was switched to intravenous metronidazole with vancomycin (both PO and enemas). Due to worsening clinical course, patient was taken to the operating room for exploratory laparotomy and noted to have markedly inflamed and dilated bowel with large-amount ascites. A diverting loop ileostomy was created and anterograde vancomycin flushes were initiated. Patient was started on total parenteral nutrition with nothing per oral status. A clear liquid diet was slowly initiated as she demonstrated ostomy function with resolution of ileus. Her diet tolerance and ostomy function improved gradually and she was transferred out of the ICU to the general medical unit on post-operative day 10.

Discussion: HCWs are at higher risk of Clostridium colitis and acute diarrheal illness in such patients should be investigated immediately and Clostridium difficile infection ruled out. Early surgical consult should be considered in patients who do not respond to conventional antibiotic therapy. A diverting loop ileostomy with intra-colonic flushes helped recovery in this patient who did not respond to medical therapy. This also preserved the colon from total colectomy which sometimes is the only option in cases of fulminant Clostridium difficile colitis.
Refractory hypoglycemia as the initial manifestation of primary empty sella

Authors: Hagop Ghareebian, MD, Amir AlDabagh, MD, Orisiman Adekolujo MD

Introduction: Empty sella syndrome (ESS) is characterized by subarachnoid space herniating into the sella turcica, causing the displacement or flattening of the pituitary gland. While primary ESS has an unknown etiology, secondary ESS may be caused by trauma, infection, adenoma, ischemia, surgery, pharmacological or radiological treatment. It has been noted in 5-23% of the population, usually found incidentally in autopsies or imaging. However, 25% to 50% of patients present with endocrine abnormalities (i.e. panhypopituitarism, diabetes insipidus, diabetes mellitus type 2, or irregular menses). Neurological (headache) or ophthalmological symptoms may also be seen.

Case Presentation: 55-year-old African American female with PMHx of anemia, presented with lethargy and acute encephalopathy after several months of progressively worsening dizziness, muscle weakness, and fatigue. For the past few days, her appetite was poor. She also complained of cold intolerance, brittle hair, dry skin, and occasional constipation. She denied headache, blurry vision, chest pain, neck pain, palpitations, or previous head imaging, trauma or surgery. Her remote history included an uncomplicated pregnancy; menopause occurred several years ago. She was found to be hypoglycemic with blood glucose 35. Six D50 ampules initially improved her hypoglycemia but her blood glucose quickly dropped thereafter. She was placed on D5 drip; with the same results. She was alert and oriented to place and person only but lacked a goiter and had trace edema. Labwork revealed low serum cortisol <1.2, high TSH 37, low free T4 0.2, low insulin 1.6, low C peptide 0.9, mildly elevated prolactin, and low FSH. UDS was unremarkable. ACTH stimulation test revealed adrenal insufficiency. She was started on IV decadron 2mg BID and her hypoglycemia resolved (later weaned to prednisone 5mg BID). In view of her hypothyroidism, adrenal insufficiency, and low FSH post-menopause, hypopituitarism was suspected. MRI brain revealed empty sella without micro- or macroadenoma.

Discussion: PES is usually asymptomatic but can present with endocrine, neurologic, or visual symptoms. A fraction of patients present with nonspecific headache, dizziness, or cranial nerve disorders. Others may have ophthalmological symptoms such as blurry or decreased vision, or mild papilledema. Though mild hyperprolactinemia and various levels of hypopituitarism have been noted, it is rare that adult patients are hospitalized for undiagnosed hypopituitarism with refractory hypoglycemia or severe hyponatremia. Panhypopituitarism is noted in only 2% of ESS patients. Our case represents a rare clinical presentation of primary ESS, with no known trauma, surgery, or ischemia (Sheehan’s syndrome). As clinicians, we should include ESS in our differential when assessing patients presenting with hypopituitarism, refractory hypoglycemia, or neurological/visual symptoms. After ruling out drug overdose, insulinemia, alcohol ingestion, infections and malignancies, MRI brain should be obtained if ESS is suspected.
Benign Uterine Leiomyoma: A Rare cause of Severe Hypercalcemia mediated by Parathyroid related peptide (PTHrp)

Authors: Sahla Hammad MD, Lauren Clever MS4, Israa Al-Gburi MD, Zarak Hassan Khan MD, Amitha Kakulavaram MD, Sandeep Garg MD

Introduction: Hypercalcemia is a relatively common clinical problem encountered routinely by primary care physicians. Among all causes of hypercalcemia, primary hyperparathyroidism and malignancy are the most common, accounting for greater than 90 percent of cases. There are some benign causes of hypercalcemia reported however they are found to be extremely rare with incidence found to be less than 10%. These include Systemic Lupus Erythematosus (SLE), Human Immunodeficiency Virus (HIV), benign tumors of the kidney and ovaries, benign pheochromocytoma.

Case Presentation: We report a case of a 41-year-old Hispanic female who presented to the emergency department with nausea and vomiting associated with her menstrual periods. On physical exam, the patient was found to have palpable uterus up to the level of the umbilicus, with no other significant finding on examination. Lab data revealed a severely high calcium level of 17 mg/dl. Serum phosphorus, sodium, potassium, chloride, creatinine was found to be within normal limits. The patient was initially managed with vigorous hydration and zoledronic acid. Pregnancy was excluded with a negative urine pregnancy test. She underwent further workup for hypercalcemia and her Parathyroid Hormone (PTH) level was found to be suppressed at 4.0 picogram/ml, 25-Hydroxy Vitamin D level was found to be 16 ng/ml. The Patient underwent diagnostic workup for multiple myeloma which was found to be negative. On further blood work, her parathyroid hormone-related peptide (PTHrp) level was elevated at 66 picogram. Bone Scan was done to rule out metastatic bone disease and came back negative. The patient further underwent a CT scan of the chest which was negative however a CT abdomen/pelvis showed a 15x15cm uterine mass consistent with suspicion of a uterine myoma. MRI abdomen was done following this which revealed a 15 x 15 cm mass and per radiological report findings were consistent with a leiomyoma or a leiomyosarcoma. The patient subsequently underwent Total Abdominal Hysterectomy (TAH) and final pathology report revealed findings consistent with a benign uterine leiomyoma. She was followed up as an outpatient following the surgery to monitor the calcium levels which were noted to be normal (9.1mg/dl) not requiring any treatment with bisphosphonates.

Discussion: The most common causes of hypercalcemia are noted to be primary hyperparathyroidism and malignancy. PTHrp related hypercalcemia is most commonly associated with malignant conditions. However PTHrp-mediated hypercalcemia associated with a benign tumor is extremely rare and has been described in uterine leiomyoma on rare occasions. This case illustrates one of the few cases of benign leiomyoma producing PTHrp causing hypercalcemia. Thus, clinicians should consider uterine leiomyoma as a potential cause of hypercalcemia mediated by PTHrp in absence of other common causes.
Multiple Liver Abscesses Caused by Streptococcus Intermedius Bacteremia in the Setting of a Routine Dental Cleaning

Authors: Angy Hanna MD, Fadi Odish MD, Zaid Imam MD, Bhavinkumar Dalal MD

Introduction: Streptococcus intermedius is a member of the Streptococcus anginosus group, also referred to as the “Streptococcus milleri” group. The S anginosus group is part of the oral cavity and GI tract normal flora. S intermedius has been known to cause abscesses in various locations in the body, most commonly in the liver and brain, however this is uncommon in healthy individuals. We report the clinical case of an immunocompetent 21 year old male found to have multiple hepatic abscesses associated with streptococcus intermedius in the setting of a recent dental cleaning and EBV infection.

Case Presentation: Patient is a 21-year old immunocompetent male who presented to the hospital with complaint of right upper quadrant pain, fevers, and chills. The patient was diagnosed with an acute serologically confirmed EBV infection 1 week prior to his admission but was otherwise healthy. He underwent a routine dental cleaning 3 months prior. On admission, his vitals were BP 88/53 mmHg, HR 142 beats/minute, RR 41 breaths/min and temperature 38.7°C. His exam was remarkable for right upper quadrant tenderness and splenomegaly. The remainder of his diagnostic evaluation revealed (a) leukocytosis of 16,000 cells/mm^3 with left shift, (b) elevated creatinine of 2.66 mg/dl, (c) elevated alkaline phosphatase of 573 U/L, aspartate aminotransferase of 102 U/L and alanine aminotransferase of 65 U/L with a bilirubin of 1.8 mg/dl. Computed-tomography (CT) of the abdomen and pelvis demonstrated multiple right lobe hepatic abscesses with the largest measuring 7.3 cm. Two sets of blood and fluid cultures from an ultrasound guided aspiration returned positive for S. Intermedius. The patient underwent 2 subsequent drainage procedures for the larger lesion. The patient demonstrated remarkable clinical recovery and was discharged with a 6-week course of PO ertapenem therapy. Follow-up laboratory values demonstrated resolution of his hematological abnormalities, acute kidney injury and transaminitis.

Discussion: This case is unique in the sense that liver abscesses in the setting of S. Intermedius typically occur in immunocompromised patients, however this case involved an otherwise healthy 21 year old recently diagnosed with EBV. To our knowledge, this is the only case involving liver abscesses occurring with a co-infection of EBV. Furthermore, S. Intermedius has been known to cause liver abscesses in the setting of dental abscesses requiring extraction, however only one other case has been reported in the literature regarding the development of liver abscesses after a routine dental cleaning. It is also remarkable that there was a 3-month interval period between the dental cleaning and initial presentation of septic shock-other cases have reported an interval period of up to 1.5 months. This case leads us to consider the potential risks associated with routine dental cleanings, even in an immunocompetent state.

References

Primary isolated hepatosplenic sarcoidosis mimicking malignancy and causing symptomatic hypercalcemia

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Introduction: Sarcoidosis is a granulomatous disease that can affect any organ system. While it is known that sarcoidosis most often affects the lung and intra-thoracic lymph nodes, it is a multisystem disease that can affect any organ. Although it is not unusual for sarcoidosis to involve the liver and spleen, it is however usually in the setting of multiorgan involvement. The presentation of sarcoidosis in one or two organ systems exclusively is quite rare, especially if it does not involve the thorax. In addition, while it is a classic finding, hypercalcemia is always an interesting finding for any sarcoidosis patient. Therefore, the rarity of our case ensues in the association of all those findings together, while mimicking malignancy early in the diagnosis.

Case Presentation: This is a case of a 67-year-old female, known to have multiple medical problems, mainly papillary thyroid cancer status post total thyroidectomy and cervical neck dissection in addition to radioactive iodine currently in remission for 1 year, presented to the hospital with severe weakness and fatigue associated with weight loss. The initial work-up showed: total calcium was high at 13.2 mg/dl (normal range: 8.7-10.4 mg/dl), ionised calcium was high at 1.5 mmole/l (normal range: 1.1-1.3 mmol/l), PTH was low at 1.7 pg/ml and her PTH-related protein was normal at 14 pg/ml (normal range: 14-27 pg/ml). The patient was treated with hydration and Pamidronate and her hypercalcemia and symptoms improved. The differential was wide, however, a CT scan of the chest, abdomen and pelvis did show multiple liver and splenic nodular lesions, with a clear chest; therefore malignancy was the highly possible diagnosis. Biopsy of the largest splenic lesion confirmed the diagnosis of sarcoidosis.

Discussion: Liver and spleen involvement in sarcoidosis has been well described, though it is almost always a part of a multisystem involvement of the illness. In 2001, a review report did show that 30% of sarcoidosis patients had 2 organ involvement, one of which was almost always in the thorax. Isolated primary hepatosplenic sarcoidosis has been reported, though it is quite rare with only a few cases reported in the literature. Isolated primary hepatosplenitis sarcoidosis has also been reported to be mimicking malignancy where sometimes it even causes elevations in tumour markers like CA-125. Hypercalcemia is a reported finding in 10% of all sarcoidosis patients. One of the most interesting findings in our reported case is the patient’s hypercalcemia that misled the diagnosis further, suggesting malignancy to be high on the differential. Hypercalcemia in isolated non-lung sarcoidosis has also been reported, but it is still quite a rare finding. Finally, a few reports have emerged where hypercalcemia in sarcoidosis was treated with bisphosphonates alone or in combination with steroids, which reflects the successful treatment of our patient with hydration and Pamidronate alone in achieving normal levels of calcemia.

References

Secondary Evans Syndrome, A Rare Presenting Manifestation of Systemic Lupus Erythematosus

Authors: Anubhav Jain, MD, Ankita Aggarwal, MD, Sarwan Kumar, MD, Wayne State University School of Medicine, Ascension Providence Rochester

Introduction: Evans Syndrome is a rare condition which is seen in 0.8–3.7% of patients with autoimmune hemolytic anemia (AIHA) or idiopathic thrombocytopenic purpura (ITP). It can present de-novo as a primary ES or may manifest as presenting features of other diseases like SLE and other lymphoproliferative disorders. There are only anecdotal reports of such cases in the literature. We present a rare case of secondary ES as the first manifestation of SLE presenting with bleeding from multiple sites and hemolytic anemia.

Case Presentation: A 24 years old female with previous normal menstrual cycles presented to gynaecology with history of intermittent spontaneous vaginal bleeding for two months and generalized weakness for six months. No obvious gynaecological cause for bleeding was apparent after preliminary workup. Thus, she was referred to the medicine department for systemic evaluation.

A detailed evaluation revealed no history of fever, weight loss, hair loss, oral ulcers, photosensitivity and arthropathic symptoms including morning stiffness. On examination she had stable vitals, conjunctival pallor with a scleral clot over right eye and petechial hemorrhages all over body. Detailed systemic examination was unremarkable. Her routine blood examination detected anemia (Hb- 4.3gm/dl), low platelet count (5x103/ µl), normal total leukocyte count and corrected reticulocyte count of 7.4%. Peripheral smear (PS) showed microcytic hypochromic anemia with spherocytes. Bone marrow (BM) aspirate was cellular with erythroid hyperplasia and megakaryocytosis. BT, CT, PT, APTT, d-dimer levels were normal. There was unconjugated hyperbilirubinemia with normal transaminases. Direct antiglobulin (Coomb’s) test was positive (3+). Anti-nuclear antibody (ANA) study was positive (2+, diffuse homogenous pattern) with a titer of 1:320, positive dsDNA (2+) with a titer of 95.0 IU/ml which was suggestive of SLE. Coagulation profile was positive for lupus anticoagulant (LA) and anti-cardiolipin antibodies. Thus, the patient was diagnosed with autoimmune hemolytic anemia and immune thrombocytopenic purpura with SLE.

The patient was given three units of whole blood on two consecutive days after admission and eight units of platelets during the hospital stay. She was also given methylprednisolone 1 gm/day as an infusion for 3 days by intravenous route and was started on tablet prednisolone 1mg/kg from the fourth day onwards. Twelve days after hospitalization, she was discharged in a relatively improved condition with a hemoglobin of 9.1 gm/dl and platelet count of 1.52 x 103 /µl on a daily dose of 1mg/kg of oral prednisolone and was advised to attend the outpatient clinic.

Discussion: Our study highlights the need for awareness of the rare entity of secondary ES. A high index of suspicion should be kept by primary care physicians, gynecologists in cases having thrombocytopenia and evidence of hemolysis to look for any underlying secondary etiologies in order to avert a misdiagnosis with attendant consequences in treatment and prognosis.

References


Delicious but Dangerous: Acute Hepatitis triggered by Cassia Cinnamon Use

Authors: Dillon Karmo M.D., Fadi Odish M.D., Ramy Mando M.D., Sandor Shoichet M.D.

Introduction: In the most recent National Health Interview Survey (NHIS), 33.2% of adults employed complementary health approaches, while 17.7% used supplements. Physicians must recognize the adverse effects of these agents. One example of this is the increasingly popular practice of using cinnamon supplementation to improve glycemic control in Type 2 Diabetes Mellitus. However, less attention has been given to a potential adverse effect of cinnamon use. Coumarin is a compound found in cinnamon (and in higher concentrations in the cassia cinnamon subtype), and its metabolite is hepatotoxic. Our patient presented with acute hepatitis triggered by the use of a seemingly-innocent supplement.

Case Presentation: A 51-year-old male presented to the Emergency Department with a chief complaint of right upper quadrant abdominal pain, jaundice, yellow colored stools, and dark urine for the past 10 days. He denied tobacco, alcohol, or illicit drug use. Medications included aspirin, allopurinol, atorvastatin, and metformin. However, his wife admitted that she has been giving him cassia cinnamon supplements without his knowledge for the past month. Physical exam was significant for scleral icterus, jaundice, abdominal distention, and tenderness to palpation in the RUQ. He had a normal CBC, Albumin = 4.3 g/dL, Alkaline phosphatase = 281 U/L, AST = 287 U/L, ALT = 906 U/L, Bilirubin, total = 5.9 mg/dL, Bilirubin, direct = 4.0 mg/dL, PT = 10.8, with INR = 1. Acute hepatitis panel, ANA, anti-smooth muscle antibody, anti-mitochondrial antibody, ferritin, HIV, HSV, EBV, and CMV were negative. An ultrasound of the abdomen was negative for any abnormalities. His atorvastatin and cinnamon supplements were held. An MRCP was obtained and revealed no evidence of biliary ductal dilation, cholelithiasis, or choledocholithiasis. His clinical status and liver function tests improved over two days, and he was discharged home with instructions to hold the atorvastatin until his liver function tests normalized, and to discontinue the cassia cinnamon.

Discussion: This case illustrates the importance of a thorough history when evaluating a patient. Special attention must be paid to over-the-counter medications, herbs, and supplements. Often times patients will not volunteer this information when asked about what medications they take, so they must be specifically prompted. Given the increasing popularity of alternative and “natural” medical practices, it is also imperative that physicians stay up to date on the potential adverse effects. Emphasis must be placed on evidence-based assessment of the risks and benefits of alternative medical practices.

References

The Bleeding Cirrhotic - Evaluation of Coagulopathic States in End Stage Liver Disease

Introduction: Coagulopathy is common in patients with cirrhosis due to an imbalance in hemostasis. Hyperfibrinolysis in the setting of cirrhosis is a condition that can present like disseminated intravascular coagulation (DIC), leading to life threatening bleeding. Although both present similarly, treatment modalities differ.

Case Presentation: A 67-year-old male with medical history of alcoholic liver cirrhosis and recently diagnosed IgA nephropathy presented to the hospital with encephalopathy and generalized weakness. Vitals on admission were within normal limits, CT head was unremarkable, and labs were normal except for a significantly elevated ammonia level. He was admitted with acute hepatic encephalopathy and treated with lactulose with improvement. During his admission, he developed left nare epistaxis for which otorhinolaryngology was consulted and initially resolved with cauterization and packing. Subsequently, however, he developed recurrent epistaxis, after which he became hypoxic requiring mechanical intubation and transfer to the intensive care unit (ICU). He continued to have epistaxis requiring packed red blood cell transfusions (pRBCs) and eventual embolization of the sphenopalatine artery. Workup was significant for elevated D-dimer, decreased fibrinogen and normal PTT and INR. Despite these values, he was given a diagnosis of severe DIC. The patient received multiple blood products, including pRBC’s, fresh frozen plasma, platelets, cryoprecipitate, and DDAVP. He developed fulminant renal failure requiring initiation of continuous renal replacement therapy. Throughout his ICU course he continued to have recurrent epistaxis, with additional development of coffee ground emesis, melena, and bleeding from his lacrimal glands and quinton catheter site. Hemodynamic instability ensued, requiring vasopressor support. He failed to improve, and in fact worsened, despite continued aggressive transfusion of blood products, leading to the eventual decision of his family to withdraw care.

Discussion: This case illustrates the importance of recognizing hyperfibrinolytic states in patients with cirrhosis presenting with massive bleeding. This process needs to be differentiated from DIC, which presents similarly. Hepatic cirrhosis results in complex changes of hemostasis due to an imbalance between anticoagulant and procoagulant factors. PT, INR, and aPTT are difficult to interpret and may not reflect increased risk of bleeding in cirrhotic patients. Previous case reports have shown that thromboelastography is useful in assessing coagulation status in those with cirrhosis, as it tests both platelet function and coagulation by assaying several parameters of clot formation in whole blood. There are also reports of other tests, including tissue plasminogen activator (t-PA), hepatic clearance of t-PA, thrombin activatable fibrinolysis inhibitor, and synthesis of alpha-2 antiplasmin and plasminogen activator inhibitor which can be used to assess hyperfibrinolytic states. The labs above, along with assessing factor VIII levels can help guide management. Hyperfibrinolysis in cirrhosis has been reported to respond well to anti-fibrinolytic agents such as tranexamic acid, and could have been considered in this patient.

References

Mitral Pseudostenosis, Severe Pulmonary Hypertension and Right Heart Failure due to Giant Cardiac Myxoma – A Mimicker

Authors: Hafiz Khan, Mahin Khan, Hameem Changezi, Ahmad Munir, Shagufta Ali

Introduction: Cardiac Myxoma (CM) are benign tumors and mostly present in the left atrium (LA)(75%) but can also be located in other chambers of the heart.1 Small CM are asymptomatic whereas giant myxoma can cause complications due to atrioventricular valve obstruction and thromboembolism for which early diagnosis is warranted.1 We present a case of CM with obstructive complications due to mitral pseudostenosis resulting in severe pulmonary hypertension (PH) and right heart failure (RHF).

Case Presentation: A 47-year-old smoker female with no cardiac history presented with worsening exertional dyspnea, malaise and weight loss for 6 weeks. She had no physical limitations 6 weeks ago but was unable to climb one flight of stairs without getting short of breath. Physical examination was pertinent for jugular venous distension, basal crackles in lungs, 2+ pedal edema and rumbling diastolic murmur at apex. She was hypotensive, tachycardiac and required 2L nasal cannula to maintain oxygen saturation. CT chest revealed pulmonary congestion, bilateral pleural effusions and hypodense filling defect in LA. She was started on supportive treatment with diuretics and oxygen supplementation. The acute coronary syndrome was ruled out by no ST-T wave changes and negative troponins. Echocardiogram showed normal EF, mild mitral regurgitation, severely increased right ventricle with decreased systolic function, severe tricuspid regurgitation, and severe pulmonary hypertension. A giant 5.5 x 4.5 cm mobile density likely myxoma was attached to the interatrial septum and prolapsing into the left ventricle during the diastolic phase causing functional mitral stenosis. Cardiac catheterization revealed no coronary artery disease. Cardiothoracic surgery was consulted who recommended surgical removal along with valve repair. She underwent resection of CM through the transseptal approach, septal reconstruction, mitral and tricuspid valves repair, and closure of left atrial appendage. The histopathology report confirmed the diagnosis of myxoma and post-operative recovery was uneventful.

Discussion: CM are benign tumors and vary in sizes between 1-15 cm in diameter.1 The clinical features are dependent on the location, size, and mobility of CM; most of which arise either due to intracardiac obstruction or thromboembolism or constitutional symptoms.1,2 In our case, CM was prolapsing into left ventricle during the diastolic phase causing functional mitral stenosis which resulted in severe PH and RHF. The size, location and interatrial connection of CM are important factors for pulmonary and systemic thromboembolic complications.

Interleukin-6 secreted by myxoma can cause constitutional symptoms including fever, malaise, arthralgia, and myalgia.3 Echocardiography is the diagnostic modality of choice to make the diagnosis and formulate management plans.4 Surgical resection is the treatment of choice for myxomas.5 Our patient underwent successful surgical resection along with valves repair. In 2%-13% of the cases, CM can recur, therefore, it is recommended to do a semi-annual follow-up for four years after resection with echocardiography.5

This case highlights a giant CM with obstructive complications and underlines the importance of early diagnosis and prompt treatment to reduce both obstructive and embolic complications.

References

A Rare Case of Spontaneous Tumor Lysis Syndrome in Metastatic Colon Adenocarcinoma

Authors: Sharmeen Mahmood, DO; Hadi Mohammed, MD; Junior Uduman MD; Ding Wang, MD

Introduction: Tumor lysis syndrome (TLS) is a life-threatening oncological emergency, characterized by metabolic derangements as outlined by the Cairo Bishop criteria. Spontaneous TLS (STLS) in the absence of any definitive treatment is a rare manifestation of solid organ tumors. Here, we report a case of metastatic colon cancer manifesting with fatal STLS. To the best of our knowledge, only two other cases of STLS in colon cancer have been reported in the literature.

Case Presentation: A 55-year-old morbidly obese male was seen by his primary care physician for two months of worsening abdominal pain and 45lb weight loss. On physical exam, a palpable abdominal mass was appreciated. A computed tomography (CT) scan of the abdomen and pelvis revealed hepatomegaly from innumerable liver masses. Liver biopsy revealed necrotic tissue necessitating a second biopsy which suggested pathologic diagnosis of metastatic adenocarcinoma of colonic origin. Subsequent colonoscopy revealed a nearly occluding, large, friable mass in the sigmoid colon. His course was complicated by lower GI bleed requiring ICU admission. Initial laboratory studies showed acute kidney injury, hyperphosphatemia, hyperkalemia, and hyperuricemia, consistent with spontaneous TLS. CEA was >8000ng/mL and Carbohydrate Antigen 19-9 was >18,000. Despite supportive care, he rapidly progressed to multiorgan failure. The patient became altered secondary to his critical illness, and his wife became his decision-maker. Given his poor prognosis, she approved transitioning to comfort care, and he expired a few hours later.

Discussion: Prophylactic treatment of TLS is clearly indicated in clinical practice for patients with hematological malignancies. Unfortunately, such precautions are rarely taken in patients with solid malignancies. This rare presentation of STLS in metastatic colon cancer highlights the need to exercise caution when managing patients presenting with bulky disease and extensive liver involvement with renal dysfunction. The initial liver biopsy for this patient revealed extensive necrotic tissue without viable tumor cells, which may have alluded underlying TLS pathophysiology. Identifying patients with risk factors for TLS, and implementing prophylactic management with fluid hydration and allopurinol based on laboratory and clinical response might avoid immediate STLS associated fatal outcomes.

References

Functional Vitamin B12 Deficiency As A Result of Nitrous Oxide Abuse

Introduction: Functional vitamin B12 deficiency is a diagnostic dilemma, with deleterious and progressive effects if missed. It can occur due to chronic nitrous oxide abuse, which eventually leads to myeloneuropathy in the setting of normal B12.

Case Presentation: A 30y.o. Female, with past medical history of polysubstance abuse and depression, presented with a three day history of ascending paresthesias and weakness in both lower limbs, along with gait instability. She reported chronic use of nitrous oxide whippets for one year, increasing over the last few months. Physical examination showed reflexes 2/4 in the bilateral biceps, triceps, brachioradialis, and absent in patellar and Achilles. Hoffman and Babinski signs were absent. Pinprick and vibratory sensations were decreased from her toes bilaterally up to her abdomen. Further evaluation revealed normal level of B12, elevated methylmalonic acid and homocysteine, along with normocytic anemia. CT head, MRI thoracic and lumbar spine, HIV, thyroid function test, protein electrophoresis, and lumbar puncture with fluid analysis were all normal. Eventually, she was diagnosed with functional vitamin B12 deficiency, leading to symptoms of subacute combined degeneration, in the setting of nitrous oxide (“laughing gas”) abuse.

Discussion: Nitrous oxide is being increasingly used as a drug of abuse by youngsters and health care practitioners. The toxic effect of nitrous oxide is mediated through oxidation of cobalt ions in vitamin B12, causing it to be deactivated. This leads to reduced conversion of homocysteine to methionine and prevents methylation of myeline proteins, causing demyelination in the central and peripheral nervous systems. The most common presentation of nitrous oxide misuse is myelopathy involving the dorsal columns, leading to subacute combined degeneration of the spinal cord. Corticospinal tracts can also be involved. However, it is very important to rule out other etiologies of neuro/myelopathy including Guillain-Barre syndrome and transverse myelitis. The mainstay of the treatment is complete cessation of nitrous oxide use. Vitamin B12 replacement can be initiated. There are no data supporting replacement of methionine. Substance abuse counseling is another area that should be focused on. Recovery from nitrous oxide induced myelopathy is slow, and a small percentage of patients have permanent deficits. Physicians should consider nitrous oxide abuse as a differential diagnosis in young patients with a history of substance abuse presenting with limb paresthesias, sensory loss and weakness. Nitrous oxide is easily available in the form of commercially available cartridges or whipped cream canisters called “whippits”.

Conclusion: In the setting of myeloneuropathy, when other differentials have been ruled out, functional B12 deficiency and nitrous oxide abuse should be on the differential, especially in high risk patients. Given the increasing availability of laughing gas at liquor stores, it is something every practitioner must be aware of.

References

The Scent of Strawberries that led to the Resolution of Hypersensitivity Pneumonitis

Introduction: Though not approved by the Food and Drug Administration for smoking cessation, e-cigarettes are frequently substituted for cigarettes, and perceived to be safe. E-cigarettes heat a liquid containing nicotine, fruity flavors, and other chemicals creating a vapor to inhale ("vaping"). Reports of pulmonary complications due to vaping include eosinophilic pneumonia, lipoid pneumonia, hypersensitivity pneumonitis, bronchiolitis obliterans (‘popcorn lung’), diffuse alveolar hemorrhage, respiratory bronchiolitis, interstitial lung disease, and acute respiratory distress syndrome. We present a case of acute hypersensitivity pneumonitis due to vaping.

Case Presentation: A 48 year old woman with nonalcoholic steatohepatitis (NASH) with transjugular intrahepatic portosystemic shunt (TIPS), and hypertension presented due to dyspnea and dry cough for a few days. She reported that she quit smoking. On examination: Afebrile, normal blood pressure, respiration rate: 24/min, heart rate: 90/min, pulse oximetry: 80%, on 3L O2: 94%. Hepatomegaly and a few bi basal crackles were found. Chest X-ray: Right lower lobe atelectasis. Complete blood count was normal, with normal differential white cell count. She was placed on oxygen and antibiotics for possible right lower lobe pneumonia. CT scan with contrast ruled out pulmonary embolism. Arterial blood gas on 3L oxygen: pH: 7.45, PaCO2: 43.1 mmHg, PaO2: 59 mmHg, A-a gradient: 115.4. Due to persistent hypoxemia, and NASH with TIPS, an echocardiogram with bubble study was done, which did not show opacification of left atrium, ruling out hepatopulmonary syndrome. Patient was spending a lot of time in the bathroom and her room smelled of strawberries. Subsequently, the nurse saw the patient vaping with strawberry flavored e-cigarette. Her CT chest was reviewed and was found to have mosaic attenuation of the lungs and air trapping, a pattern seen in hypersensitivity pneumonitis. Patient was told to stop vaping and was started on methylprednisolone. She improved dramatically with resolution of hypoxemia and was discharged on steroid taper.

Discussion: Hypersensitivity pneumonitis occurs due to inhalation of a variety of organic antigens leading to an inflammatory reaction that presents as an acute, subacute or chronic condition. Acute hypersensitivity pneumonitis presents with dry cough, dyspnea, fever, and hypoxemia. Leukocytosis may occur, but eosinophilia is not seen. Chest X-ray frequently shows fine reticulonodular pattern but can be normal. High resolution CT scan can show ground glass opacities, mosaic attenuation on inspiratory images and air trapping on expiratory images. Removal of the offending agent is the most effective therapy. In patients with persistent or progressive symptoms, steroids are effective. Chronic hypersensitivity pneumonitis presents as pulmonary fibrosis and does not respond to medical therapy. Since early removal of the offending agent is the mainstay of treatment, detailed exposure history is important. Hypersensitivity pneumonitis should be considered in patients who smoke e-cigarettes, and vaping should be stopped to improve outcome.

References

A rare fatal case of Salmonella aortitis diagnosed with positron emission computed tomography scan

Authors: Sheda Monfared Beheshti, MD. James Passinault, MD.

Introduction: Salmonella aortitis (SA) is a rare rapidly progressive infection of the aorta with fatal endovascular complications, most commonly diagnosed with contrast enhancing computed tomography (CT). It often presents with non-specific symptoms of fever, chills, and fatigue, making time-sensitive diagnosis and treatment challenging. Left untreated it will lead to rupture, with more than 90% mortality rate.

Case Presentation: An 83-year-old Caucasian male with past medical history of poorly controlled diabetes and a 3.5 cm abdominal aortic aneurysm (AAA) presented to the emergency department with three days history of chills, fatigue, and worsening shortness of breath. On presentation, he was febrile, tachypneic, and tachycardiac. Physical examination was remarkable for diffuse fine crackles on lung auscultation and bilateral lower extremity pitting edema. Laboratory workup was remarkable for neutrophilic leukocytosis. Initial blood cultures grew Salmonella Dublin, and treatment with IV Cefepime was initiated. CT abdomen/pelvis with contrast showed a stable 3.5 cm abdominal aortic aneurysm with the identification of a new 3.3 cm left iliac aneurysm with no evidence of endovascular infection. Repeat blood cultures continued to grow Salmonella Dublin from an unknown source of infection. Follow up transthoracic echocardiogram, and transesophageal echocardiogram were unremarkable. Due to high suspicion for infectious aortitis in the setting of Salmonella Dublin bacteremia and a history of a pre-existing aortic aneurysm; a whole body positron emission tomography (PET) CT was ordered that showed infection of the aortic wall. Infectious disease recommended a 12 weeks course of IV antibiotics as the patient was not a surgical candidate secondary to multiple comorbidities and overall deconditioning. At two weeks follow up repeat CT abdomen/pelvis with contrast showed worsening of soft tissue density of both the AAA and the iliac artery mycotic aneurysm with the formation of a new infrarenal mycotic aneurysm. At thirty days follow up the patient was severely deconditioned, unable to stand independently, with a significant limitation of mobility. At three months follow up shortly after completing a 12 weeks course of IV antibiotics, the patient had recurrent Salmonella bacteremia with extensive damage to the aortic, iliac, and femoral arteries; sadly his care was transitioned to comfort care.

Discussion: Salmonella aortitis (SA) is a rare fatal cause of a mycotic aneurysm in patients with pre-existing atherosclerotic disease and diabetes. The general clinical picture is one of delayed diagnosis and high mortality. A high index of suspicion is required when treating the high-risk population with persistent bacteremia and fever of unknown origin despite aggressive IV antibiotics and negative contrast enhancing CT imaging. Therefore, awareness of this rare yet fatal clinical finding and the value of alternative diagnostic imaging including PET CT scan is required to ensure early diagnosis, timely treatment, and improved patient survival.
Glimepiride-Induced Autoimmune Hepatitis

Authors: Ali Mrad M.D., Nimmish Khera, M.D., Katherine Doyle D.O., Niluka Weerakoon M.D.

Introduction: Glimepiride is a second-generation sulfonylurea used in the management of non-insulin-dependent diabetes mellitus. Per our review of the literature, few cases were reported about hepatic injury due to glimepiride use. We present a case of autoimmune hepatitis associated with the use of glimepiride in a patient with no previous history of a hepatobiliary disease.

Case Presentation: A 77-year-old male presented with chief complaint of severe pruritus with yellowish discoloration of his skin and eyes for one month. Patient has a medical history of Transient Ischemic Attack, Essential Hypertension, Type 2 non-insulin dependent Diabetes Mellitus, and Gastroesophageal Reflux Disease. There is no history of a hepatobiliary disease, blood transfusion, alcohol or illicit drug use. The patient was on Plavix, Losartan, Metformin, Glimepiride, and Prilosec. Physical exam was positive for jaundice, and scleral icterus, but otherwise negative. There was no abdominal tenderness, guarding or distention. Initial testing revealed normal white blood cell and platelet count. Aminotransferase (ALT) and Aspartate aminotransferase (AST) were elevated, with elevated total and free bilirubin, and alkaline phosphatase. Hepatitis A, B, C, and E testing was negative. Acetaminophen level was normal. Iron studies were normal, not indicating hemochromatosis. Carcinoembryonic Antigen (CEA), Carbohydrate antigen (CA) 19-9, α-fetoprotein, α-1 Antitrypsin levels were normal. Mitochondrial Antibody, Liv-Kid Micro Antibody, P-ANCA were negative. The antinuclear antibody (ANA) and anti-smooth muscle antibody level came back positive. Abdominal ultrasound showed Coarsened hepatic parenchyma, which may correlate for hepatocellular disease with no biliary dilatation. Computed tomography for abdomen and pelvis, and Magnetic resonance cholangiopancreatography (MRCP) revealed no significant findings in the hepatic biliary tract. Based on that, the etiology thought to be Autoimmune Hepatitis. The patient was also instructed to hold the glimepiride since there were concerns of possible medication-induced hepatobiliary injury as was reported in the literature. Patient's symptoms improved significantly right after. Three months after discontinuing the glimepiride, repeated antinuclear antibody and anti-smooth muscle antibody were strikingly negative.

Discussion: This case illustrates the possible hepatic injury induced by glimepiride use. That injury was easily reversible after discontinuing the insulting medication. Clinicians need to aware of that possibility of liver injury associated with the use of glimepiride. Baseline liver function tests should be obtained before starting the medication. Patients with a history of hepatobiliary disease should have their liver function test assessed periodically. If there is any rise of the liver function tests, Glimepiride should be discontinued immediately.
A Rare Case of Atrio-Esophageal Fistula Post Atrial Fibrillation Ablation.

Authors: Ahmad Murad M.D., Manar Barakat M.D., Asaad Nakhleh M.D.

Introduction: Atrioesophageal fistula (AEF) is a very rare complication of atrial fibrillation (AF) and radiofrequency ablation (REA). It was described for the first time in two very experienced centers in 2004, and it is the most dreadful and lethal among all the others related to REA. Its clinical presentation is extremely variable, and data on management is conflicting and at the present, there is no consensus on the most effective treatment strategy for AEF. Although the incidence is less than 0.1%, it is usually fatal. The high case-fatality rate has traditionally been attributed to the lack of recognition and late presentation of this complication.

Case Presentation: We present a case of a 42-year-old male with a past medical history of AF on anticoagulation who underwent left atrial RFA and pulmonary vein isolation and presented with confusion and bilateral upper and lower extremity weakness.

The patient tolerated the procedure but complained of difficulty swallowing afterwards. Five days later, he began having malaise and GI upset which was associated with an episode of loose bright red blood BM. He later became more confused, began to have weakness in all of his extremities, and was admitted to the hospital for workup. Imaging of the brain revealed multiple focal areas of subacute infarcts consistent with multiple embolic events involving the bilateral frontal lobes, left parietal and right temporal lobes. CT of the chest also revealed air next to the posterior wall of left atrium which was thought to be secondary to AEF as a complication of his RFA.

His hospital course was complicated by seizure, sepsis secondary to aspiration pneumonia, and cardiac arrest requiring several rounds of chest compressions. Immediately after he regained pulse, he was noted to be posturing with worse mental status, and a CT of the head showed multiple air emboli bilaterally with a higher stroke burden when compared to his initial CT scan. The patient was deemed non-operative, and was eventually discharged to long term acute care facility.

Discussion: Our patient was a young male who had an AEF as a complication of RFA for AF. The patient’s AEF was diagnosed after he presented with an embolic stroke, and was managed conservatively. Our case is unique due to the fact that CPR caused worsening in the fistulous communication between the left atria and the esophagus leading to higher burden of embolic strokes. This case emphasizes the importance of early diagnosis and treatment of AEF as worse outcomes can be seen if recognition of this complication is late.
Dissecting an Interesting Case of Chest Pain in a Young Healthy Mother

Authors: Rohan Naik, MD, Hongfeng Yu, MD, PhD, Omar Elmadhoun, MD, MPH, John Haapaniemi, DO, Sabeeh Siddiqui, MD, Camelia Arsene, MD, PhD

Introduction: Spontaneous coronary artery dissection (SCAD) is a non-traumatic, non-iatrogenic, and non-atherosclerotic separation of the coronary arterial wall. SCAD has emerged as an increasingly recognized cause of acute coronary syndrome (ACS) and sudden cardiac death, particularly in younger patients and in women who lack conventional atherosclerotic risk factors. SCAD reportedly accounts for 1-4% of all ACS cases and 35% of myocardial infarctions in women ≤50 years of age. The left anterior descending (LAD) artery is the most commonly affected artery by this phenomenon.

Case Presentation: A 32-year-old healthy female recently delivered a baby uneventfully. On post partum day 9, she developed acute chest pain and extremity edema. She went to a local hospital and was found to have elevated blood pressure and mildly elevated troponin I (0.23 ng/mL). Left heart catheterization (LHC) was performed showing no coronary artery blockage. She was discharged with Aspirin, Atorvastatin, Metoprolol, Lisinopril and Furosemide. Her edema improved, however, the chest pain worsened. She presented to our hospital on postpartum day 12 with complaints of severe chest pain radiating to her back. She had elevated serial troponin I of 1.02 ng/ml, 8.53 ng/ml and 14.66 ng/ml, which eventually peaked at 32.07 ng/mL. Electrocardiogram (ECG) showed inverted T waves in V5-V6, which were not evident on previous ECG’s. Echocardiogram revealed a reduced ejection fraction of 35-40%, along with findings of severe hypokinesis in the inferior-lateral wall and apex. She received emergent LHC, which revealed extensive dissection of mid to distal left anterior descending and proximal to distal left circumflex coronary arteries. She was treated with heparin infusion for 72 hours to prevent progression of thrombosis and maintain coronary perfusion. Dual Anti-platelet Therapy was also added.

Discussion: Postpartum coronary artery dissection is an important cause of acute myocardial infarction in young females. The female sex, labor and delivery, fibromuscular dysplasia, connective tissue disorders, and valsalva type activities are some risk factors.

Diagnosis and management of acute SCAD can be challenging and requires a high index of suspicion as well as familiarity with the condition. Early Coronary angiography, Intravascular ultrasound (IVUS) and Optical coherence tomography (OCT) are invaluable in confirming the diagnosis.

Conservative medical therapy is indicated if the patient is clinically stable and does not have high risk coronary anatomy. This often results in complete resolution of dissection. For clinically stable patients with isolated left main dissection, urgent Percutaneous coronary intervention (PCI) is indicated. Coronary artery bypass grafting (CABG) should be considered in patients with Left main SCAD progressing to involve the LAD and/or proximal 2-vessel coronary artery dissection. If an isolated LAD dissection progresses to a ≥ proximal 2-vessel coronary artery dissection, conservative therapy may be reasonable. For SCAD with acute ongoing ischemia or hemodynamic instability, consider PCI or urgent CABG, although high risk

References

Organizing Pneumonia is not just a COP-out Diagnosis

Authors: Logeswari Neelakandan, Jared I.Tucker, Michael E.Nissan, Apoorva V.Saoji, E M Malitha S.Hettiarachchi, Thomas J.Piskorowski

Introduction: Cryptogenic organizing pneumonia (COP), formerly known as bronchiolitis obliterans organizing pneumonia (BOOP), is a well documented but rare condition characterized by idiopathic deposition of fibroblasts and myofibroblasts compromising the terminal bronchioles and alveoli. As part of a chronic inflammatory process, the disease progresses through a series of stages beginning with alveolar epithelial injury leading to infiltration of inflammatory cells. In progression, fibroblasts replace the inflammatory cells which manifest as the accumulation of fibrin, a process denoted by the name “organizing pneumonia.” Clinically, COP presents as progressive shortness of breath in a patient with or without constitutional symptoms. Radiographically, COP most commonly projects as nonspecific, patchy ground glass opacities. Consequently, an extended investigation almost exclusively supersedes failed antibiotic response, leading to delays in diagnosis. We present a case of atypical COP with diffuse nodular opacities.

Case Presentation: A 57-year-old African American male presents with progressive dyspnea and predominant nonproductive cough for 3 months, further associated with diaphoresis, chills, and unintentional weight loss. He concedes a history of prolonged incarceration, parakeet handling, chronic tobacco and marijuana use. He discloses having sought medical attention near the onset of symptoms at which time chronic obstructive pulmonary disease (COPD) was diagnosed, radiographically. Clinically, this is a nontoxic middle-age appearing patient with right basilar inspiratory crackles, lacking compelling extrapulmonary findings. Initial chest radiograph showed innumerable diffuse interstitial pulmonary nodules, concerning for miliary tuberculosis. There was no systemic inflammatory response syndrome criteria met. Eosinophilia was, notably, absent. Serial AFB and sputum cultures were negative. Urine drug screen demonstrated cannabinoids. HIV antigen-antibody assay was negative rendering opportunistic infections unlikely. In addition to urine Legionella antigen, Mycoplasma pneumoniae, Chlamydiae pneumoniae, and Chlamydiae psittaci IgM were negative. Beta-D glucan, Aspergillus, and Histoplasma antibody were negative. High-resolution CT scan demonstrated diffuse, tiny pulmonary nodules delineated in a bronchovascular distribution. Albeit ANA titer positivity (1:1280), ANCA was negative making vasculitis unlikely in this presentation, further implying the need for Bronchoscopy. Bronchoalveolar lavage cultures were unyielding.

Bronchoscopy biopsy indicated acute on chronic inflammation with interstitial fibrosis; meanwhile, lacking evidence for granuloma formation, haemorrhage, lymphoma and epithelial malignancy. Subsequent biopsy via video-assisted thoracoscopic surgery (VATS) exhibited polypoid plugs of organized spindle cells with collagenous stroma situated within the lumens of distal airways and peribronchial airspaces, without other explained pathological processes.

Discussion: Declaring a diagnosis of COP requires clinical, radiographic and histopathological features mutually consistent with obstructive pneumonia; simultaneously, lacking evidence for all other conceivable etiologies. Radiographically, this presentation demonstrates innumerable diminutive nodules arranged in a diffuse bronchovascular pattern – a distinct deviation from the classically described patchy ground glass opacities. Given the nonspecific symptoms of COP, it is imperative to diligently pursue the diagnosis in order to initiate prompt therapy, accurately prognosticate, and minimize sequelae.
Severe Pneumocystis jirovecii Pneumonia during treatment for Cushing’s Syndrome

Authors: Zaid A Noori MD, Rebecca Witherell, MD and Joel T Fishbain MD, FACP

Introduction: *Pneumocystis jirovecii* pneumonia (PCP) was a rare opportunistic infection until the AIDS epidemic. PCP in non-HIV patients develops in patients receiving high dose steroids, various chemotherapeutic and immunosuppressive agents. We report a case of severe PCP during therapy for Cushing’s syndrome.

Case Presentation: A 67 year old female was recently diagnosed with an ACTH producing carcinoid tumor of the lung. Treatment was initiated with mifepristone as an outpatient but ketoconazole was started as an inpatient. She was initially admitted for diabetic ketoacidosis but two weeks after starting the ketoconazole she developed fevers, respiratory distress, hypoxia and rapidly progressive pulmonary infiltrates on chest x-ray. Chest CT imaging demonstrated extensive ground-glass opacities. Based on her history, a very high LDH level and deteriorating course she was empirically placed on high dose sulfamethoxazole/trimethoprim. A diagnosis of PCP was confirmed by a markedly elevated beta-D-glucan and positive sputum PCR for *Pneumocystis jirovecii*. She developed acute respiratory distress syndrome requiring a prolonged course of intubation. She was subsequently extubated and transferred to a long term acute care facility.

Discussion: Unlike HIV patients who present with a more indolent course, non-HIV patients often have a rapidly progressive course. The most common time frame for patients on high dose steroids to develop symptoms is during the steroid taper. Our patient presented during treatment for endogenous production of cortisol with documented lower levels of cortisol. Early recognition and therapy is critical as this condition results in significant morbidity and mortality.
Role of C peptide while switching from Insulin to Oral Hypoglycemics in Type 2 Diabetes Mellitus

Authors: Vinisha Noti, Kavitha Kesari

Introduction: C-peptide levels reflect function of the β cells of pancreas in Diabetes. C-peptide levels are associated with diabetes type and duration of the disease more so with type 2 DM. C-peptide levels less than 0.08nmol/l confirms absolute insulin deficiency and the definitive need for insulin to prevent complications. (Normal fasting level is 0.3–0.6 nmol/l; postprandial level increases to 1–3 nmol/l) There have been several advances in assays that have made C-peptide measurement both reliable and inexpensive. Despite this, measurement of this hormone in the clinical settings is rarely done. We present a case of type 2 DM who was inadvertently switched from Insulin to oral hypoglycemic agent and readmitted with Hyperosmolar Hyperglycemic state(HHS) complication.

Case Presentation: A 91- year-old male with history of well controlled Type 2 Diabetes Mellitus (Baseline HbA1c 6.6) on Insulin for the last 30 years was admitted to the hospital for CHF exacerbation. During the hospital course, he was found to have a HbA1c of 5.9. The physician in charge of the patient, discharged him on glipizide instead of insulin. The Patient presented to the hospital 3 weeks later in HHS, with blood sugars above 600mg/dl and in sepsis secondary to urinary tract Infection. His repeat HbA1c was 9.3 and fasting C- peptide level was low at 0.34ng/ml (0.81-3.85 ng/ml) equivalent to 0.10nmol/l. He was treated with Insulin, IV fluids and antibiotics. He was later switched to insulin and his blood sugars were well controlled.

Discussion: In type2 DM, there is a progressive decline in C-peptide level that correlates with decreased endogenous insulin secretion capacity. Treatment with Oral hypoglycemic agents (OHAs) acting through enhancement of beta cell insulin secretion would appear to have less response in those who do not secrete endogenous insulin. This explains our patient’s scenario with low C-peptide levels. Switching the patient to glipizide from Insulin led to glycemic instability as reflected by precipitation into HHS and high HbA1C. The high demand metabolic state caused by sepsis unmasked and precipitated the existing insulin deficiency into absolute deficiency leading into HHS in our patient

In majority of the clinical scenarios, the decision to initiate OHAs and or Insulin is based on glycemic control status or HBA1c. However, while switching a patient from Insulin to OHAs, HbA1C alone may not be a strong predictor for the successful maintenance of OHAs. Patients with type 2 DM who have preserved beta cell function might be more likely to be maintained on oral hypoglycemic agents after switching from initial insulin therapy. In this regard when considering insulin withdrawal or when considering addition of OHAs dependent on endogenous insulin, c-peptide could be used as a predictor of pancreatic beta cell function in patients with type 2 diabetes.

References

Twiddler doo dah, twiddler eh! My oh my the shocks are in my armpit!

Authors: Olusola Ogundipe, MD; Geetha Krishnamoorthy, MD, FACP

Introduction: The implantable cardioverter defibrillator (ICD) devices have late complication rates of 2 to 3% and only 4% of those who suffer late complications experience lead dislodgement. This complication presents as perforation, arrhythmia or inappropriate pacing of adjacent structures. We present a case of Twiddler syndrome, a rare complication, where there is dislodgement of ICD leads from its original location, with subsequent stimulation of adjacent nerves.

Case Presentation: A 54 year old gentleman with atrial fibrillation and ischemic cardiomyopathy who had a dual function ICD/pacemaker inserted 1 year prior to current hospitalization, presented to us with perceivably irregular and rapid heartbeats. In addition he reported twitching, paresthesia and electric shock-like sensation when lying on his left side. He had been diagnosed and treated at various times as possible neuropathy, myoclonus and focal seizures without relief. On examination, he was hemodynamically stable, with an irregularly irregular heart rate on auscultation, at a rate of about 140 beats/min. His ICD pocket appeared normal. Electrocardiogram showed atrial fibrillation with rapid ventricular response without any identifiable pacemaker spikes. Chest x-ray identified the device in the left chest, however initial report failed to identify its malposition. Close inspection of chest radiograph and comparison to previous studies revealed the device had migrated downwards from its original insertion pocket and the lead which was coursing just inferior to the left clavicle and into the right atrium on the old chest x-ray was now wrapped around the ICD generator. Electrophysiology consult was sought for device interrogation and reimplantation.

Discussion: Twiddler syndrome is a rare cause of ICD malfunction that can result from deliberate or accidental manipulation of the pulse generator within its pocket. This results in dislodgement of its lead(s), stoppage of ventricular pacing, and subsequent sensing of myopotentials from adjacent structures, usually the phrenic nerve, that causes pacing of the diaphragm. Patient may also experience pulsation in the abdomen. Dislodgement, may also lead to stimulation of brachial plexus that causes twitching of the left arm in a rhythmic fashion. Careful evaluation of Chest x-ray can quickly identify lead malposition, reducing associated morbidity and the risk of sudden cardiac death from ICD malfunction. Patients with ICD who report new localized abnormal muscle twitching in the chest, neck or upper limbs, sensation of abdominal pulsation or recurrent palpitations could have lead dislodgment secondary to Twiddler syndrome. Physicians should obtain a Chest x-ray to look for lead dislodgement.
Candida Krusei: How Did It Get There?

Authors: Sara Samaan, MD; Leonard B. Johnson, MD, FACP

Introduction: Candida krusei is an uncommonly isolated Candida species that is increasing in prevalence. However, it is a very rare cause of empyema. We present a case of empyema due to Candida krusei.

Case Presentation: 79 year old female with history of hypertension, coronary artery disease, and hyperlipidemia presented with worsening dyspnea for three hours. She was recently hospitalized for two days after choking on a piece of chicken at home. Endoscopic evaluation revealed a hiatal hernia. On examination, the patient was tachypneic, hypotensive, and afebrile, with diminished breath sounds in the left lower lung, extending two-thirds of the way up the chest. Chest imaging revealed a large left loculated pleural effusion with compressive atelectasis of the left lung. Initial WBC was 14200. The patient was admitted and empirically treated with Ampicillin-Sulbactum for suspected aspiration pneumonia, and possible empyema. She initially underwent thoracentesis, followed by video-assisted thoracoscopic surgery with chest tube drainage. Pleural fluid cytology was negative for malignancy, but the culture grew Streptococcus mitis oralis, Streptococcus salivarius, and Candida krusei. This prompted the addition of Anidulafungin to the treatment regimen. A subsequent swallow evaluation did not reveal a leak from the esophagus or the stomach. On day 12, she had dark stools, and her hemoglobin decreased from 11.7 to 6.4. This prompted an upper endoscopy which revealed an 8 mm esophageal fistula within the hiatal hernia at the gastroesophageal junction. The patient underwent laparoscopic repair of the fistula and was continued on Ceftriaxone and Anidulafungin.

Discussion: This is a rare case of Candida krusei empyema, with only four cases previously reported in the literature. Candida species usually colonize mucous membranes and the digestive tract, so thoracic involvement typically results from an esophageal perforation, or fistula. In the absence of known risk factors, a diagnosis of Candida species in the pleural space should prompt upper gastrointestinal evaluation. Of note, non-albicans Candida species have varying susceptibilities to antifungal agents; Candida krusei is known to be fluconazole-resistant, but is susceptible to Anidulafungin, Amphotericin, and the newer azoles.

References


Uncovering a Hidden Cause for Recurrent Diarrhea

Authors: Rovin Saxena MD, Mayuri Kulkarni MD, Gurinder Pal Gakhal MD, Cecilia Cosma MD

Introduction: Clostridium difficile infection (CDI) has become a common cause of recurrent diarrhea. Up to 35% of patients experience recurrent infection after initial therapy. Patients with recurrent CDI are occasionally treated empirically with metronidazole or vancomycin. CDI is a burden due to the increase in incidence of recurrence over recent years. In fact, there is a 50-60% chance of a repeat recurrence. Persistent diarrhea is also commonly caused by microscopic colitis, which has an incidence of 7.9 per 100,000 person years. It has been associated with the use of non-steroidal anti-inflammatory agents and proton-pump inhibitors (PPI). We present a case of a patient with recurrent CDI who was eventually diagnosed with microscopic colitis.

Case Presentation: A 41-year-old male with a history for diabetes mellitus type 1, renal failure, and gastroesophageal reflux disease presented with persistent diarrhea for the past 5 years. His temperature was 98.3 degrees fahrenheit, heart rate 87 bpm, blood pressure 123/89 mmHg, and RR was 20 per minute. Physical exam was unremarkable including his abdominal exam. Laboratory studies were insignificant. Computed tomography with contrast was negative for any pathologies. Initial stool testing was negative for CDI. A colonoscopy was performed which was negative for anatomic abnormalities or masses. Biopsy of the colonic mucosa was normal. He was treated with IV fluids and loperamide. His symptoms persisted and repeat stool was positive for Clostridium difficile toxin. He was started on metronidazole and his PPI was discontinued. His symptoms resolved over 48-72 hours and he was discharged. Two weeks later, he presented again with CDI and was treated with oral vancomycin and discharged. He presented with another recurrence of diarrhea 2 months later and was treated empirically for possible CDI with metronidazole, showing with minimal to no improvement of his symptoms. Stool studies were negative for Clostridium difficile toxin. Patient underwent a repeat colonoscopy and biopsy which showed lymphocytic infiltrates within the colonic mucosa confirming the diagnosis of lymphocytic microscopic colitis. He was started on budesonide with a good clinical response and was discharged. Follow-up at our outpatient clinic showed near-resolution of his symptoms.

Discussion: Recurrent diarrhea is bothersome and can decrease a patient’s quality of life. Since the patient presented in this case had a past medical history of CDI-induced diarrhea, microscopic colitis was overshadowed. Our patient was treated empirically for CDI diarrhea, putting him through unnecessary exposure to antibiotics and delaying the appropriate treatment. Antibiotic usage may create iatrogenic consequences for the patient, such as antibiotic resistance. Clinicians must be aware that patients with history of recurrent CDI may have other underlying etiologies for persistence of diarrhea.

References

MICHIGAN CLINICAL VIGNETTE POSTER FINALIST - REHANA SIDDQUI, MD

A Rare Case of Endogenous Endophthalmitis arising from Fulminant MRSA Bacteremia

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Introduction: Endogenous endophthalmitis (EE) is a rare, potentially-blinding, ocular infection caused by hematogenous microbial seeding of the posterior eye segment. This report presents a rare case of EE arising from fulminant MRSA bacteremia.

Case Presentation: A 66-year-old female with history of cataract surgery and chronic back pain presented with generalized septic arthritis and blurred vision. She had previously undergone pedicle screw fixation with L2-L6 fusion, complicated by MRSA bacteremia, for which she subsequently underwent revision surgery and completed an 8-week course of vancomycin four months prior to presentation. On admission, she was found to have recurrence of MRSA bacteremia. Examination revealed Roth spots, vitreous debris, and diffuse retinal hemorrhages, consistent with bacterial EE. Administration of intravitreal vancomycin and moxifloxacin ophthalmic resulted in complete resolution of visual symptoms. Further workup revealed spinal hardware loosening, epidural abscess, aortic and mitral valve endocarditis, septic pulmonary emboli, splenic abscess, and septic cerebrovascular emboli that resulted in ischemic strokes. Blood cultures were successfully cleared with removal of spinal hardware, abscess drainage, and treatment with daptomycin, linezolid and ceftaroline.

Discussion: The incidence of EE is 1 in 50,000, and the majority of cases arise from fungemia. Although rare, it is critical to remain vigilant for EE when visual symptoms accompany bacteremia, as EE does not respond to intravenous antibiotics, and can progress to permanent blindness. There are multiple known risk factors, and particularly in this case, development of endocarditis from bacteremia significantly increased the patient's risk of EE, as it is the leading cause of EE in the United States.
Think Outside The Lung!

Authors: Zahra'a Salah, MD, FACP ; Amit Mohindra, MD, FACP ; Ramesh Mohindra, MD, FACP

Introduction: Meigs' Syndrome consists of a triad of benign ovarian tumor (commonly a fibroma), pleural effusion and ascites. Ovarian Remnant Syndrome is a condition in which physiologically active ovarian tissue is inadvertently left behind post bilateral salpingo-oophorectomy. Meigs' Syndrome and Ovarian Remnant Syndrome are rare conditions, with Meigs' Syndrome occurring in less than 1% of ovarian tumors. This is a unique case of a woman who, although had total abdominal hysterectomy with bilateral salpingo-oophorectomy in the past, developed Meigs' Syndrome due to having Ovarian Remnant Syndrome.

Case Presentation: The patient was a 79 year old female with past medical history of hypertension, stroke, and recurrent pleural effusions resulting in multiple hospitalizations. Past surgical history was remarkable for total abdominal hysterectomy with bilateral salpingo-oophorectomy at the age of 40 for contraception. She underwent thoracentesis five times. Each time, 1000-1400 ml of exudative fluid was removed, with no evidence of infection or malignancy. During her last hospital admission, physical exam showed decreased breath sounds and dullness to percussion in the left mid-lower lung fields. Abdominal exam showed mild distention with no hepatosplenomegaly. Chest x-ray showed left sided pleural effusion. Pleural fluid analysis was unremarkable. Chest CT scan showed no mass but did reveal abnormal hepatic texture. Abdominal ultrasound was performed revealing a retroperitoneal mass. CT scan of the abdomen demonstrated a 15x12 cm retroperitoneal mass with ascites. Biopsy of the mass was consistent with an ovarian fibroma. Later testing for cancer antigen 125 (CA-125) level was mildly elevated.

Discussion: Reviewing this case proved that diagnosing Meigs' Syndrome, presenting with only signs and symptoms of pleural effusion, can be difficult. It becomes even more challenging, to have Meigs' Syndrome in mind as a differential diagnosis, in a patient who has had surgical history of total abdominal hysterectomy with bilateral salpingo-oophorectomy. The importance of diagnosing both Meigs' Syndrome and Ovarian Remnant Syndrome is that they can be effectively treated with surgical removal of the ovarian tissue and the prognosis for MS is excellent with resolution of the pleural effusion and ascites postoperatively.

References

Angiovac: A novel approach to treat Endocarditis

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Introduction: Angiovac is a suction thrombectomy device approved for percutaneous removal of clots or tumors from the intravascular system. We describe a patient with tricuspid valve endocarditis and multiple pulmonary emboli who was not a surgical candidate. He successfully underwent resection of a large tricuspid valve vegetation using the Angiovac system.

Case Presentation: A 45-year-old intravenous heroin abusing man presented with dyspnea. A Chest CT revealed multiple infiltrates consistent with septic emboli. A transthoracic echocardiogram demonstrated a 5.5cm tricuspid valve vegetation. Blood cultures remained positive for MSSA despite escalating antibiotics to daptomycin and nafcillin. Considering the size of the vegetation and persistent bacteremia, surgery was contemplated. The patient was deemed a poor surgical candidate due to his hemodynamic instability and underlying multiple comorbidities. AngioVac suction of the vegetation was offered to the family. The patient tolerated the procedure well. Repeat blood cultures were negative. Clinically he improved and was transferred to the LTAC facility.

Discussion: The tricuspid valve is involved in 10-25% of all cases of endocarditis. Antibiotics remain the cornerstone of treatment. Surgical intervention may be indicated for 1) persistent bacteremia 2) recurrent embolization despite appropriate antibiotics, 3) large vegetations (more than 20 mm) or 4) refractory congestive heart failure. Due to the patient’s poor nutritional status, respiratory failure due to multiple septic emboli and sepsis, he was deemed an unsuitable candidate for open cardiac surgery. The use of the novel Angiovac system may be considered for tricuspid vegetation resection in endocarditis patient who are poor surgical candidates.
The Crystal Crown.

Authors: Hasan Ahmad Hasan Albitar, Department of Internal Medicine; Mayo Clinic, Rochester, MN., Floranne Ernste, Division of Rheumatology; Mayo Clinic Rochester, MN.

Introduction: Crowned dens syndrome (CDS) is a rare cause of febrile neck pain. It should be suspected in patients with acute neck pain (with or without a history of CPPD arthropathy) and can be confirmed radiographically (calcium deposits around dens) as highlighted by our case below

Case Presentation: A 74 year old man with a history of AML presented with right shoulder and neck pain and fever of 2 days duration. Physical examination revealed temperature of 39 Celsius and right shoulder tenderness. Workup revealed leukopenia with absolute neutrophil count of 0.29 x10(9)/L. Head and cervical vertebrae CT scans were negative. He was started on Vancomycin and Cefepime for neutropenic fever. Later, this was deescalated to prophylactic antimicrobials as the infectious workup was negative.

Subsequently, the patient developed new bilateral foot pain and worsening shoulder pain. ESR was 130 mm/hr, CRP was 23.1 mg/L, and uric acid was 3.4 mg/dL. Right shoulder arthrocentesis showed rhomboid shaped and positive birefringent crystals consistent with CPPD crystals. Dual energy CT scan showed diffuse calcium deposition about both ankle joints and mid feet consistent with CPPD deposition. Given the above findings; our suspicion for CDS was high. We reviewed the cervical spine CT scan obtained on admission and calcifications of the transverse ligament were detected which confirmed the diagnosis of CDS. The patient was started on 40 mg prednisone with complete resolution of his symptoms.

Discussion: CDS is a rare syndrome characterized by severe neck pain, fever; and elevated inflammatory markers. CT scan is the gold standard for diagnosis; the key radiographic finding is the presence of peri-odontoid calcification in a crown or halo configuration. CDS has a favorable response to treatment with oral NSAIDs, colchicine, and corticosteroids. High index of suspicion is required to diagnose CDS as it can be frequently missed on imaging.

References

Hidden in Plain Sight: Vertebral Artery Dissection

Authors: Shravya Vinnakota MBBS, Korosh Sharain MD

Introduction: Vertebrobasilar insufficiency often presents with subtle signs that require a high index of suspicion for accurate and timely diagnosis. Clinical features can include local symptoms like headache, neck pain or focal neurological deficits; however, these are often mild or absent.

Case Presentation: A 57-year-old male presented with complaints of progressively worsening nausea and vertigo for 3 weeks. This was associated with intermittent headaches recurring every few days. Prior to symptom onset, he slipped on ice and landed on his back, causing a whiplash injury. He denies direct trauma to the head or loss of consciousness. He had no improvement in symptoms with Ibuprofen. His past medical history is significant for sensory seizures secondary to a benign intracranial dysembryoplastic tumor, well-controlled on Levetiracetam for several years. Physical exam revealed right-sided horizontal nystagmus. A non-contrast head CT did not reveal any acute pathology. His nausea and vomiting improved with conservative measures. However, he continued to have intermittent vertigo, therefore, an MRI with angiography of the head and neck was obtained and revealed a right vertebral artery dissection (VAD) and acute infarction of the right cerebellar hemisphere. He was initiated on daily Aspirin and high-intensity statin therapy. CT Angiography of the abdomen/pelvis was obtained to look for associated conditions and was negative. The patient was discharged with outpatient neurology follow-up. His symptoms gradually resolved and he underwent surveillance imaging after 3 and 6 months, which demonstrated a stable evolving infarct.

Discussion: Spontaneous artery dissections are uncommon but account for 25% of strokes in the young. Etiology can be varied and difficult to identify. Trauma of varying degree, especially in the setting of underlying risk factors, has been linked to VAD. Subsequent ischemia is more common than hemorrhage as it is thought that emboli from thrombus formation at the dissection site are the cause for the ischemic sequelae. Recent literature suggests a 3-step bedside oculomotor exam (HINTS-head impulse test, nystagmus and test of skew) is more sensitive than early MRI for ischemic strokes. Non-contrast head CT has only 16% sensitivity for detecting acute ischemia. The diagnosis is made by neuroimaging with CT angiography or MRI with angiography of the head and neck. In the setting of ischemia, thrombolytics are indicated in the initial 4.5-6 hours. Additionally, antithrombotic therapy with antiplatelet/ anticoagulant drugs is recommended for at least 6 months. Endovascular interventions or surgery are considered for patients with subarachnoid hemorrhage or recurrent ischemic symptoms while on antithrombotic therapy. Secondary preventive measures should target cardiovascular risk factors. For an internist, it is crucial to consider vascular dissections in the differential for headache and vertigo in a young adult, especially with any trauma to the head.
Don’t Monkey Around with Antibiotics

Introduction: Baboon syndrome is a systemic contact dermatitis that occurs when a patient previously sensitized to a contact allergen is exposed to the same allergen through a systemic route and develops a well demarcated erythematous patch at the axillae, upper inner thighs and buttocks. When there is no known history of previous cutaneous sensitization, this reaction is termed systemic drug-related intertriginous and flexural exanthema (SDRIFE).

Case Presentation: 62 year old man with aortic valve regurgitation status post bioprosthetic aortic valve replacement and a recent diagnosis of prosthetic valve Streptococcus mitis infective endocarditis, presented to the hospital with daily fever, malaise and myalgia two weeks into his intravenous (IV) ceftriaxone therapy. He also noted that his face and chest were pruritic after antibiotic infusion. He denied any skin changes. He reported a history of penicillin allergy but had a negative skin testing two years ago. He recalled developing a pruritic rash in his groin when he was given oral penicillin in his early twenties. On exam, he was non-toxic appearing, hemodynamically stable and had a grade 3/6 systolic murmur heart best at right upper sternal border unchanged from previous exam. Lab showed leukopenia with white blood cell count 1.6 x 10^9/L (3.5-10.5 x 10^9/L) with absolute neutrophil count of 830 x 10^6/L. Differential diagnosis for his fever included failed antibiotic therapy versus adverse reaction from ceftriaxone. His IV ceftriaxone was switched to IV penicillin G in the setting of possible drug reaction and leukopenia. Within hours after receiving IV penicillin G, he developed a symmetric, erythematosus and pruritic patch in his groin and bilateral inner thighs without systemic symptoms. He was evaluated by an allergy specialist, and his history and exam was consistent with SDRIFE. Patient was recommended to avoid penicillins. He was switched to IV daptomycin and completed total six weeks of IV antibiotic therapy.

Discussion: Skin complaints are commonly encountered in both outpatient and inpatient, and given the frequent use of antibiotics, it is important that clinicians recognize possible side effects including Baboon syndrome and SDRIFE. Baboon syndrome or systemic contact dermatitis is a delayed type hypersensitivity reaction by non-cutaneous exposure to an allergen in a patient who is previously cutaneously sensitized. The characteristic rash is a well-defined symmetrical erythema of the gluteal area and in the intertriginous folds without systemic symptoms. The time between allergen exposure and onset of dermatitis ranges from hours to a few days. When there is no known previous cutaneous exposure, it is diagnosed as SDRIFE. SDRIFE is most commonly associated with beta-lactam antibiotic and has a male predominance. It is a self-limiting condition that can be treated symptomatically with topical or oral antihistamine and steroid. Baboon syndrome and SDRIFE should be considered in patients presenting with intertriginous rash.

References

A 37 year-old woman with lower extremity claudication

Authors: Caitrin M. Coffey, Uma Thanarajasingam

Introduction: Lower extremity claudication is a common complaint seen by the general internist, but an uncommon presenting symptom in large vessel vasculitis. Early recognition of key clinical features of Takayasu arteritis is essential to avoid further vascular damage in a young patient with claudication symptoms.

Case Presentation: A 37 year-old Caucasian woman presented for evaluation of right foot tingling and exertional right leg pain that was relieved with rest of 2 months’ duration. Her medical history was notable for congestive heart failure diagnosed after an episode of viral myocarditis two years prior. She underwent cardiac catheterization during that time which did not show coronary disease, and suffered left renal artery thrombosis causing renal infarction shortly thereafter, prompting left nephrectomy. Medications included pravastatin, bisoprolol, lisinopril, bumetanide, potassium, aspirin, and levothyroxine. Review of systems was positive for neck pain, shoulder stiffness, and negative for chest pain, shortness of breath, edema, and fevers.

Upon presentation, blood pressure was 110/55 in the left arm. Right arm pressure was unable to be obtained as the brachial pulse could not be auscultated. Physical examination revealed left carotid bruit, diminished radial pulses, non-palpable pedal pulses, and lower extremity varicosities. Cardiac and pulmonary auscultation, skin, and joint examinations were normal.

Laboratory testing included ESR 28, CRP 32.3, NT-pro-BNP 1045 pg/mL, creatinine 1.3, ANA 0.4, anti-RNP IgG 1.1 (<1.0 negative), and negative rheumatoid factor, anti-CCP, MPO, PR3, cryoglobulins, and antibodies to double stranded DNA, SSA/Ro, SSB/La, Smith, Scl-70, and Jo-1. Ultrasound showed mild symmetric wall thickening of the common carotids bilaterally, wall thickening of the right subclavian artery, and focal stenosis in the left subclavian artery. CT angiogram showed minimal arterial wall thickening in the abdominal aorta extending to the right common and internal iliac arteries, and luminal irregularity of the aorta and right external iliac artery. PET/CT showed hypermetabolic activity in the right iliac bifurcation and in the left common carotid artery, compatible with active vasculitis.

Discussion: Takayasu arteritis (TA) is a rare large vessel vasculitis characterized by granulomatous inflammation of the vascular wall and predominantly affecting the aorta and its major branches in young women. While a majority present with upper extremity symptoms, 18% are reported to present with lower extremity claudication. In this case, this patient under 40 years of age presented with classic clinical findings including difference in pulses between the arms, unobtainable blood pressure, history of limb pain and fatigability, neck pain, and elevated ESR. Conventional angiography is considered the gold standard for diagnosis of TA, however CTA and MRA can also be used to demonstrate vessel wall thickening, mural inflammation, stenosis, and aneurysms that may occur in the disease process. Initial treatment includes high dose corticosteroids and often other immunosuppressive agents such as methotrexate, cyclophosphamide, azathioprine, and biologic agents.

References

Systemic Lupus Erythematosus in the Elderly: An Atypical Presentation of a Common Disease

Authors: Dame Idossa, M.D., Melissa Lyle, M.D., Joseph Grande, M.D., Ph.D

Introduction: Pericardial effusion is seen in more than 50% of patients with systemic lupus erythematosus (SLE) [1]. However myopericarditis, pericardial effusion, and heart failure, as the initial presentation of SLE is rare. Cardiovascular disease in SLE has a high morbidity and mortality, thus early detection and treatment are paramount.

Case Presentation: A 75-year-old woman presented with progressive dyspnea, orthopnea, lower extremity edema, pleuritic chest pain, and generalized fatigue. Laboratory workup was notable for a WBC 11.8, INR 1.8, Na 120, lactate 6, BNP 4138, and elevated liver enzymes. Electrocardiogram was notable for low voltage and mild diffuse ST elevations. Troponins were elevated, without a significant delta. Chest X-ray showed large bilateral effusions. Echocardiogram showed moderate to large circumferential pericardial effusion and mild extrinsic compression of the right atrium and right ventricle, concerning for early signs of cardiac tamponade. Pericardiocentesis removed exudative pericardial fluid. In the evaluation for hyponatremia, urine studies revealed microscopic hematuria, hyaline casts, and an elevated albumin/cr ratio, concerning for a glomerular disease process.

Rheumatological work-up revealed elevated ESR to 57, CRP of 78.9, TSH 4.7, elevated TPO 101.4, decreased C3 and C4 at <3 and 40 respectively, positive ANA, markedly elevated dsDNA of >1000, and antibodies toward SSA/Ro, SSB/La, Smith, and RNP. Electrophoresis showed polyclonal hypergammaglobulinemia. Renal biopsy revealed mesangial lupus glomerulonephritis. She was treated with immunosuppressive therapy--prednisone and Cellcept, with significant improvement in her inflammatory markers, auto antibodies, and microscopic hematuria.

Discussion: SLE in the elderly is uncommon and rarely reported with disease onset at age 65 and older [2]. In addition, large pericardial effusions and myopericarditis are also uncommon initial presentations of SLE. When present, they are often associated with nephritis and myocardial dysfunction [3]. The timely recognition and early treatment with immunosuppressive agents is imperative to prevent the mortality associated with this condition.

References

Mixed Connective Tissue Disease with Mixed Etiology Interstitial Lung Disease

Authors: Brenden S. Ingraham, MD and Marcia R. Venegas Pont, MD

Introduction: Adalimumab is an uncommon cause of interstitial lung disease (ILD). Patients on adalimumab for connective tissue disease can have a mixed ILD with fibrotic changes in addition to the ground glass opacities (GGOs) resulting from adalimumab.

Case Presentation: 64-year-old female is admitted for workup and management of subacute, progressive dyspnea, pleuritic chest pain, dry cough, and new oxygen requirement. History is significant for plaque psoriasis on adalimumab (initiated 3 months prior to presentation), ulcerative colitis status post proctocolectomy, sicca symptoms, Raynaud’s phenomenon, GERD, maternal history of rheumatoid arthritis, and a 50-pack-year smoking history. CT showed moderate, multifocal GGOs consistent with nonspecific interstitial pneumonia (NSIP) or usual interstitial pneumonia (UIP) without honeycombing. Pulmonary function testing showed severely reduced DLCO. Ejection fraction was 60% without significant echocardiographic abnormalities. Labs remarkable for normal leukocyte count; moderately elevated ESR and CRP; and mildly elevated RNP, LDH, and aldolase. Rheumatologic and infectious testing (including bronchoalveolar lavage) was otherwise negative. Transbronchial biopsy showed bronchiolar wall and parenchyma without neoplasm or granuloma. Video capillaroscopy confirmed secondary Raynaud’s. Adalimumab was stopped, and she was started on prednisone 30 mg daily. She improved clinically and was discharged with home oxygen and plans for outpatient rheumatology and pulmonology follow up. Repeat CT chest one month later showed significant improvement in GGOs but persistence of fibrotic ILD. Pulmonology believed the remaining disease represented UIP versus NSIP related to underlying connective tissue disease. Rheumatology felt that the ILD with elevated RNP in the setting of her other rheumatologic symptoms were suggestive of mixed connective tissue disease (MCTD). Azathioprine was started with plans to wean the prednisone. She has since been improving clinically.

Discussion: Tumor necrosis factor inhibitors, like adalimumab, have been implicated in rare cases of interstitial pneumonia with a prevalence of 0.5% to 3%. ILD occurs in > 50% of those with MCTD. Careful consideration should be given to the underlying cause as adalimumab-induced ILD is a diagnosis of exclusion after ruling out infectious and cardiac etiologies. It is important to recognize fibrotic lung disease independent of the GGOs from adalimumab, especially when ILD persists despite discontinuation of the offending agent. Removing the adalimumab is often sufficient, but corticosteroids can be utilized in more severe or refractory cases. Fibrotic changes in the setting of connective tissue disease may require prednisone or other steroid-sparing agents, like azathioprine.

References

Antiphospholipid Syndrome as a cause of severe gastrointestinal bleed

Authors: Sae Jang, Shounak Majumder

Introduction: Antiphospholipid syndrome is an autoimmune hypercoagulable disorder characterized by thromboembolic events. We present a case of severe acute anemia due to gastrointestinal hemorrhage.

Case Presentation: A 55 year-old woman with past history of antiphospholipid syndrome complicated by multiple deep vein thromboses and miscarriages, prior hemorrhagic stroke, chronic slow GI bleed with iron deficiency anemia, thrombocytopenia, and CKD stage 3 presents to the ED with two-month history of progressively worsening shortness of breath, dizziness, and chest discomfort with exertion, which acutely worsened over the last day. Initial evaluation revealed hemoglobin of 3.9 grams/dl and presence of heme-positive stools. Patient denied abdominal pain, hematochezia, or maroon-colored stools. She endorsed black tarry stools which she attributed to her oral iron supplementation.

She was admitted to the hospital and received 5 units of packed red blood cells with increase of Hgb to 9.7. Anticoagulation was held. EGD showed erosive gastropathy and colonoscopy showed non-bleeding plaque type lesions which were treated with argon plasma coagulation. Capsule endoscopy and Meckel’s scan were unremarkable. When patient was trialed back on anticoagulation, her Hgb dropped to 7.4.

Her hospitalization was further complicated by AKI on CKD with a peak creatinine of 2.4 and nephrotic range 24-hour proteinuria of 4.5 grams without casts. Serologic work up included positive ANA 1:160 in a speckled pattern, rheumatoid factor, and low C3 and C4. Antiphospholipid antibodies were positive. Other vasculitis and rheumatologic laboratories were negative.

Kidney biopsy showed acute renal microangiopathy with severe tubular atrophy and interstitial fibrosis. Severe hypertensive arteriosclerosis and acute tubular injury was also present. Complete repeat endoscopic work-up showed gastric antral vascular ectasia and significant segmental mucosal ulceration suggestive of ischemia. CT chest showed a wedge-shaped area of infarct in the right middle lobe.

Given her history of APS, the microangiopathy seen on renal biopsy, wedge-shaped pulmonary infarct, and ischemic-appearing colonic ulcerations, it was felt that patient’s overall presentation and severe GI bleed was due to ischemic colitis from micro-infarcts and antiphospholipid syndrome. Patient received a course of IV methylprednisolone, plasmapheresis, and rituximab infusion. She was bridged to a therapeutic level of warfarin with stable hemoglobin prior to discharge from the hospital.

Discussion: Abdominal manifestations of APS are uncommon but can be life threatening. Reported cases of thromboses include Budd-Chiari syndrome, hepatic-veno-occlusive disease and occlusion of small hepatic veins, or hepatic infarction. Intestinal manifestations are even more infrequent. This patient had catastrophic antiphospholipid syndrome defined by 1) Widespread thrombosis in multiple organ systems, 2) development of manifestations simultaneously within a week, 3) confirmation of small-vessel occlusion on histopathology, and 4) laboratory confirmation of antiphospholipid antibodies. This case highlights that abdominal involvement should be considered in patients with APS.

References

Tracheobronchomalacia: An Asthma Mimic Hiding In Plain Sight

Authors: Charles Meade MD, Kaiser Lim MD

Introduction: A 49 year old female was admitted to the medicine floors with shortness of breath. She carried a chart history of poorly controlled severe persistent asthma, as well as obesity with a BMI of 38.

Case Presentation: She described ongoing shortness of breath following recent hospital discharge. She increased use of her home non-invasive ventilation with incomplete symptomatic relief. Denied infectious symptoms and had no known history of heart failure, appearing euvolemic on exam.

In the 6 months prior to admission she had 10 ED visits for shortness of breath, and 2 additional hospital admissions. Discharge diagnoses included asthma, anxiety, and vocal cord dysfunction. Her pulmonary regimen at admission included prednisone 20 mg daily as well as an inhaled long acting beta agonist, high dose inhaled corticosteroids, and a leukotriene receptor antagonist. She had been on oral corticosteroids for most of the prior 2 years.

Non-hypoxic on presentation and arterial blood gas pH 7.46, pCO2 30, PO2 112. Prior blood gases showed similar respiratory alkalosis suggestive of hyperventilation. PFTs taken prior to admission consistently showed no reduction in FEV1/FEC ratio, no evidence of restrictive disease, no gas exchange abnormality.

Given her non-obstructive PFTs and her apparent lack of response to near-maximal asthma therapies, suspicion was high for an asthma mimic. Review of prior CT imaging was suggestive of dynamic large-airway collapse and during hospital admission she underwent limited bronchoscopy. Significant expiratory dynamic airway collapse was observed suggestive of tracheobronchomalacia. Her positive pressure support was uptitrated with good response and she was discharged with a plan for further pulmonology follow up for procedural consideration and to guide de-escalation of asthma therapies.

Discussion: Tracheobronchomalacia (TBM) involves weakening of the supporting structures of the large airways. Exhalation results in excessive dynamic airway collapse (EDAC). The epidemiology and underlying causes of TBM are poorly characterized but when significant can result in nonspecific respiratory complaints which often mimic more common pulmonary diseases such as asthma. Direct bronchoscopic observation of airway collapse is the current diagnostic gold standard although dynamic CT imaging during inspiratory and expiratory phase can also be useful.

This case highlights the usefulness of maintaining high clinical suspicion for alternative diagnosis in patients with atypical asthma presentations, patients who appear to have severe disease, and patients who are treatment resistant. TBM/EDAC is a known asthma mimic that can sometimes be identified on prior cross sectional imaging. Informative diagnostic testing is available either via bronchoscopy or dedicated inspiratory and expiratory cross sectional imaging. TBM will not respond to traditional asthma therapies and definitive treatment is often procedural.
Chronic Lymphopathy: A case of recurrent pleural effusions, lymphedema, and yellow nails

Authors: Bryan J. Neth, M.D., Ph.D., Michael Richter, M.D., Jose R. Castellanos, B.A., Marc Greenberg, B.S., and Dennis Regan, M.D.

Introduction: Yellow Nail Syndrome is a rare disorder characterized by the constellation of lymphedema, pulmonary pathology, and yellow nails. Less than 400 cases have been reported with an estimated prevalence of <1/1,000,000. Although the cause is unknown, it is thought to be related to systemic lymphatic dysfunction.

Case Presentation: An 86-year-old Caucasian man presented to the hospital with bilateral lower extremity edema and generalized weakness. The patient’s edema and weakness progressed over the last several months and had been associated with functional decline and worsening dyspnea on exertion. He had no changes in medications or recent illnesses over this time period. He denied chest pain, headache, abdominal pain, or changes in bowel or bladder. Physical examination was remarkable for 2+ pitting edema in the bilateral lower extremities extending proximal to the knees, discolored nails on his bilateral hands and feet, heart was regular rate and rhythm, and lungs were clear with diminished breath sounds at the bases. CBC and metabolic panel were within normal limits. CXR revealed bilateral (L>R) pleural effusion without noticeable pulmonary consolidation.

The patient underwent a thoracentesis, which yielded 825 mL of exudative fluid with elevated triglycerides. We next focused on diuresis (6.5 L total) with leg wraps to improve lower extremity edema. The patient’s symptoms were likely related to his underlying Yellow Nail Syndrome. The progressive nature of his symptoms over several months was consistent with prior episodes. After symptomatic management, he was discharged to a skilled nursing facility for acute rehabilitation.

Discussion: The three defining features of Yellow Nail Syndrome are lymphedema, pulmonary pathology, and yellow nails. Lymphedema most often occurs in the lower extremities and is clinically indistinguishable from primary lymphedema. The pulmonary findings can be diverse. Commonly associated pathology includes chronic cough, recurrent pneumonias, recurrent pleural effusions (95% exudative), and bronchiectasis. Yellow nails are the main clinical manifestation. Yellow Nail Syndrome has no cure. Pulmonary findings and lymphedema are treated symptomatically.

Our patient’s clinical course began with recurrent pleural effusions about 20 years prior to presentation. This necessitated thoracenteses every 4-6 months for management of dyspnea. About 3 years after his initial pleural effusion our patient began to develop extensive bilateral lower extremity and groin edema. Finally, about 10 years after initial pleural effusion our patient’s nails began to harden and eventually became discolored to a near-yellowish hue with several of his nails falling off and failing to regrow.

Yellow Nail Syndrome should be considered in the setting of lymphedema and recurrent pleural effusions, particularly if characteristic nail changes are evident. Increased awareness and research concerning Yellow Nail Syndrome could lead to improved management options that may be applicable to other disorders of lymphatic dysfunction.

References

Idiopathic Central Diabetes Insipidus, Xanthelasmas, and Bone Pain

Authors: Gordon J. Ruan, MD,1,2 Gaurav Goyal, MBBS,2 Mithun Vinod Shah, MD, PhD,2 Ronald S. Go, MD2, 1. Department of Internal Medicine, Mayo Clinic, Rochester, MN, 2. Division of Hematology, Mayo Clinic, Rochester, MN

Introduction: Erdheim-Chester Disease (ECD) is a non-Langerhans cell histiocytosis that is diagnosed histologically by foamy histiocytes demonstrating CD68+CD1a- marker on immunohistochemistry. ECD affects many organs, including the long bones, pituitary gland, cardiopulmonary vascular system, skin, kidneys, and retroperitoneum [1]. Patients often go years without a diagnosis (average of 4.2 years after initial presentation) and the prognosis is poor if not treated [2]. However, once the diagnosis is made, the prognosis is favorable as overall mortality has improved with the use of targeted therapy such as BRAF and MEK inhibitors [3-4].

Case Presentation: We present a 33-year-old lady who initially presented with polydipsia and polyuria seven years prior to her diagnosis of ECD. She was diagnosed with idiopathic central diabetes insipidus (CDI) and empirically treated with desmopressin. Five years later, she had a magnetic resonance imaging (MRI) of the brain performed after a near syncopal event. The MRI showed a 1.2-cm enhancing lesion in the hypothalamus. She underwent craniotomy and biopsy of the pituitary mass, which showed inflammatory tissue but no malignancy; repeat pituitary biopsy showed the same results. Six years after her diagnosis of idiopathic CDI, she developed many xanthelasmas in the periorbital areas bilaterally and began complaining of fatigue, intermittent nausea and vomiting, and bone pain in her lower legs and right forearm. She had x-rays of the legs that showed medullary sclerosis in the distal tibia and fibula. MRI of the left ankle showed diffuse marrow signal abnormality seen within the distal tibia/fibula and patchy lesions in the bones of the hindfoot, midfoot, and forefoot. This led to a consideration for the diagnosis of ECD and a referral was made to a hematologist. Whole-body positron emission tomography-computed tomography scan was performed that showed multiple fludeoxyglucose (FDG) avid uptake in the occipital bone, left clavicle, right and left distal radius and ulna, both femurs, both tibias, both fibulas, L5, scattered bones of the feet, and abnormal FDG uptake in the hypothalamus. Bone biopsy of the left lower leg showed nonspecific findings of sclerosis and fibrosis. Finally, a biopsy of her xanthelasma was performed and showed foamy histiocytes that were CD68+CD1a- and BRAFV600E positive on immunohistochemistry. The patient was ultimately treated with dabrafenib and trametinib and had clinical and radiologic response.

Discussion: ECD is an often overlooked diagnosis due to its rarity and heterogeneous clinical presentations and manifestations [5]. Patients often see multiple providers and undergo multiple biopsies before a diagnosis or referral to a hematologist is made. Increasing awareness about the disease is necessary as prognosis is favorable with appropriate therapy [2]. All clinicians should include ECD in their differential if they see a patient who has a constellation of idiopathic CDI, constitutional symptoms, bone pain, xanthelasmas, and/or neurologic symptoms.

References

“I thought it would make me feel better”: Altered Mental Status and Acute Kidney Injury Secondary to Hypercalcemia

Authors: David Sanborn, MD1; Catherine D. Zhang, MD1; Jimmy Mao, MD1; Keahi Horowitz1; Jason Szostek, MD1, 1Department of Medicine, Mayo Clinic, Rochester, MN

Introduction: While primary hyperparathyroidism and malignancy account for over 90% of hypercalcemia cases, it is important to consider other less common causes in symptomatic hypercalcemia that lead to hospitalization.1

Case Presentation: A 68-year-old man with hypertension, obstructive sleep apnea, chronic back and hip pain, and stage IIIA chronic kidney disease presented to the emergency department with six months of progressively worsening altered mental status with acute worsening over two weeks. CT of the head was acquired and was unremarkable. Initial laboratory studies revealed a serum total calcium of 13.1 mg/dL (8.8 – 10.2 mg/dL), serum albumin of 3.5 g/dL (3.5 – 5.0 g/dL), and serum creatinine of 5.18 mg/dL (0.74 – 1.35 mg/dL) up from 1.5 mg/dL in 2017. Upon admission, history was limited by patient’s somnolence, slow responses, and unreliability. Further workup revealed a phosphorous of 5.9 mg/dL (2.5 – 4.5 mg/dL), undetectable levels of parathyroid hormone, a 25-hydroxy vitamin D3 level of 141 ng/mL (< 80 ng/mL), and a parathyroid hormone-related peptide of 1.1 pmol/L (< 2.0 pmol/L). Serum and urine protein electrophoresis with immunofixation and angiotensin-converting enzyme levels were within normal limits. X-rays of the lumbar spine and hips were negative for lytic lesions. Intravenous (IV) lactated Ringers were started at 200 cc/hr along with calcitonin. On this regimen, the patient’s serum calcium fell to 10.5 mg/dL within 36 hours and his altered mental status improved significantly. On further questioning, he reported taking 100,000-200,000 International Units (IU) of Vitamin D intermittently over the past several months for depression. IV fluids were discontinued to attempt an oral hydration trial; however, serum calcium increased to 11.5 mg/dL and were re-initiated. Several attempts failed, and endocrinology was consulted. Both prednisone 40 mg by mouth daily and denosumab 60 mg IV x1 were administered. With this regimen, the patient was eventually able to transition successfully form IV fluids to oral hydration. In spite of maintaining calcium levels less than 11 mg/mL, creatinine remained high upon discharge at 4.53 mg/dL. Ultimately, he was discharged in stable condition with serum calcium of 10.8 mg/dL and close outpatient follow up with primary care and nephrology.

Discussion: The Institute of Medicine recommends taking no more than 4000 IU of vitamin D daily, however, more patients are taking high doses of vitamin D now than in the past.2,3,4 Because vitamin D is lipophilic, the serum availability of its various metabolites can be elevated for months resulting in an elevated serum calcium for some time.5 Treatment for hypervitaminosis D-induced hypercalcemia includes decreasing dietary intake of calcium and vitamin D, fluid hydration, and glucocorticoids.6 Further research is needed in order to better understand the effect of bisphosphonate and denosumab on hypervitaminosis D induced hypercalcemia.7

References

MINNESOTA CLINICAL VIGNETTE POSTER FINALIST - ANDREA SITEK, MD

A Rare Cause of Testicular Pain

Authors: Andrea Sitek, M.D., Kenneth Warrington, M.D.

Introduction: ANCA-associated vasculitis rarely presents with testicular symptoms.

Case Presentation: A 54-year-old man with no significant past medical history developed testicular pain one month prior to presentation. This was followed by daily fevers, night sweats, myalgias and 9 kg weight loss. He was initially evaluated in the outpatient setting and was prescribed antibiotics for presumed epididymitis with no improvement in symptoms. He was subsequently hospitalized, and an extensive infectious workup, including blood cultures and testing for Lyme disease, syphilis, tuberculosis, Legionella, Brucella, West Nile, and HIV was negative. Additional studies included bone marrow biopsy, cross-sectional chest and abdominal imaging, colonoscopy and tumor markers, all of which were unrevealing. He was ultimately discharged with a course of broad spectrum antibiotics and antifungals with arrangements to continue workup as an outpatient.

He presented to the ED one week after discharge with ongoing fevers and progression of testicular pain. Laboratory studies revealed significant anemia, leukocytosis, elevated creatinine, hematuria with renal epithelial cells and proteinuria. Testicular ultrasound showed innumerable, small hypoechoic lesions throughout both testes without Doppler flow. CT abdomen/pelvis showed mildly edematous kidneys with stranding. He was admitted to a tertiary care hospital, and Infectious Disease and Rheumatology were consulted.

A thorough infectious workup was negative. Markers of inflammation were elevated and c-ANCA was positive at 1:2048, with PR3 antibody of >8 (normal <0.4). In view of progressive kidney injury, he underwent renal biopsy. This showed pauci-immune necrotizing and crescentic glomerulonephritis, consistent with a diagnosis of granulomatosis with polyangiitis (GPA). Treatment consisted of IV methylprednisolone followed by a prednisone taper and rituximab infusions. His renal function improved, and fevers and testicular pain resolved.

Discussion: Small vessel vasculitis, like GPA, is a rare cause of testicular pain. Among patients with vasculitis, testicular pain is more often seen with polyarteritis nodosa. Our patient presented primarily with testicular pain as an initial manifestation of systemic small vessel vasculitis, an atypical presentation which resulted in delayed diagnosis and progression to kidney injury. A final diagnosis was made after identifying PR3-ANCA positivity and pursuing renal biopsy. This case highlights that vasculitis should be considered in patients with constitutional symptoms and a persistent inflammatory state. It is also important to consider that vasculitis can cause testicular pain. This can be a difficult diagnosis given the nonspecific sonographic testicular findings.
A Case of Abdominal Pain and Worsening Confusion in a 76-Year-Old Female

Authors: Elida Voth, MD, Bradly A. Kimbrough, MD, Jason Szostek, MD

Introduction: Stercoral colitis is a rare complication of chronic constipation with an estimated 200 cases reported in the literature. The hypothesized pathogenesis is that fecal impaction exerts pressure on the colonic wall and impairs transmural perfusion causing ischemia. This acute condition can rapidly progress to bowel perforation and hemodynamic instability if not promptly recognized. Clinically, stercoral colitis may present as an acute abdomen, however, the variability in physical exam and laboratory findings make it a diagnostic challenge. We describe a case of an elderly woman presenting with abdominal pain and worsening confusion in the setting of chronic constipation.

Case Presentation: A 76-year-old female presented to the emergency department (ED) with abdominal pain and worsening confusion. She had a past medical history of dementia, coronary artery disease, paroxysmal atrial fibrillation, and type 2 diabetes mellitus. Two weeks prior, she presented to the ED for worsening confusion, falls and diarrhea. Physical exam revealed lower abdominal tenderness. A CT abdomen/pelvis at that time showed a large amount of stool in the colon and rectal distention. She was discharged from the ED and instructed to increase her home bowel regimen. On the day of admission, which was 11 days later, the patient returned to the ED with copious diarrhea, several days of decreased oral intake, and a fall at her skilled nursing facility.

In the ED, the patient appeared lethargic and disoriented. On physical exam, the abdomen was soft, diffusely tender to palpation in all four quadrants, and without rebound tenderness. Rectal exam revealed copious brown stool from the rectum and soft brown stool in vault. Repeat CT abdomen/pelvis demonstrated large volume stool in the rectosigmoid colon with diffuse circumferential wall thickening, new surrounding fat stranding and vascular engorgement, and new mild mesenteric edema, all of which supported the diagnosis of stercoral colitis. Labs were significant for leukocytosis with neutrophilic shift, elevated lactate, and acute kidney injury.

Several hours after admission to medicine, there was an acute change in the patient’s abdominal exam with exquisite tenderness to palpation and new rebound tenderness. General surgery was consulted, and the patient was taken to the operating room. Abdominal exploration revealed a retroperitoneal bowel perforation due to stercoral ulcer. Unfortunately, the patient passed away several days later due to complications of the perforation.

Discussion: We report the case of a patient with chronic constipation which progressed to stercoral colitis and eventually bowel perforation. This case highlights the importance of a rectal exam and close outpatient follow-up in a patient that presents with lower abdominal tenderness and excessive stool burden on imaging. Lastly, this case reinforces the utility of CT scan and the high degree of clinical suspicion necessary in order to diagnosis stercoral colitis.
**Fever Without a Cause: An Unusual Case of Fevers and Hearing Loss**

Authors: Zachary Yetmar, M.D.; Daniel Childs, M.D.

**Introduction:** Fever of unknown origin was originally defined as a fever of 38.3°C or greater on several occasions for at least three weeks without a clear source after one week of inpatient evaluation. While no longer requiring evaluation to take place inpatient, patients with unexplained fever can present significant diagnostic challenge. Leading etiologies include systemic rheumatic disease, infection, and malignancy.

**Case Presentation:** A 35 year-old female with a history of rheumatoid arthritis on prednisone and rituximab presents for evaluation of recurrent fevers. Her symptoms started 6 months prior when she began experiencing a decline in functioning along with a 38 pound weight loss. Other symptoms include dyspnea on exertion, cough, lower extremity edema, and diffuse joint swelling. She had recurrent fevers starting 4 months ago. She was evaluated at an outside facility and was ultimately diagnosed with viral meningitis. She also had two episodes of pancreatitis without a known cause. Around this time, she experienced progressive, complete sensorineural hearing loss.

Laboratory studies included complete blood count showed lymphopenia (0.56 x 109 cells/L) without CD20 cells (0%), elevated C-reactive protein (23.5 mg/L), elevated hemoglobin A1c (7.1%), elevated cyclic citrullinated peptide antibody (186.1 units), negative antinuclear antibody (less than 1:80), hypogammaglobulinemia including immunoglobulin A (57 mg/dL), immunoglobulin M (27 mg/dL), and immunoglobulin G (650 mg/dL). Lumbar puncture was performed showing elevated total nucleated cells (35 cells/mcl), elevated protein (83 mg/dL), normal glucose (40 mg/dL), and negative paraneoplastic autoantibody panel.

Further infectious workup was also negative including human immunodeficiency virus screening, hepatitis B serology, hepatitis C antibody, Histoplasma antibody, Blastomyces antibody, Sporothrix antibody, (1,3)-beta-D-glucan assay, interferon-gamma release assay, JC virus DNA, and blood and cerebral spinal fluid cultures for bacteria, fungi, and mycobacterium.

Positron emission tomography was performed, showing diffuse inflammation throughout nearly all soft tissues and organs. A punch and muscle biopsy was performed on her left lateral thigh. Punch biopsy pathology showed septal edema and lobular panniculitis with necrosis and chronic lymphohistiocytic inflammation. Muscle biopsy showed inflammatory muscle disease. Enterovirus PCR of the muscle biopsy was positive for Coxsackievirus A9, confirming a diagnosis of disseminated enterovirus. The patient was started on intravenous immunoglobulin G, with symptomatic improvement.

**Discussion:** With advances in technology and diagnostic approach, greater numbers of patients with fever of unknown origin are able to be successfully diagnosed with their underlying disorder. A complete history and physical is the first and most essential step in evaluation. Next is a full laboratory and radiographic workup, particularly driven by features in the history. In patients who remain without a diagnosis, positron electron tomography can also be helpful in localizing pathology for more focused workup. For patient with disseminated enterovirus, there is no established standard therapy but intravenous immunoglobulin G may have benefit.

**References**

A Case of Jelly Belly

Authors: Rebecca Browning, DO; Spencer Hatch, DO; Bharat Malhotra, MD

Introduction: Ascites is known to have many causes, including cirrhosis, nephrotic syndromes, Budd-Chiari, myxedema, infections, and malignancy. The SAAG and total protein of the ascitic fluid is used to guide the diagnosis of the underlying condition. This case illustrates the importance of trusting your clinical judgement and a multidisciplinary approach to achieving a diagnosis.

Case Presentation: A 62-year-old female with a past medical history of Hepatitis C Cirrhosis, Hypothyroidism, Chronic Lymphocytic Leukemia presented with frequent falls due to her large abdomen. She reported progressively worsening abdominal girth over a one-year period that was now associated with early satiety, mixed constipation and diarrhea but denied jaundice, changes in sleep pattern, nausea, vomiting, weight loss, or night sweats. Upon arrival, exam was notable for no jaundice or scleral icterus, profoundly distended abdomen with a positive fluid wave and few scattered spider angioma.

Several paracentesis were performed. The largest was the initial paracentesis, with 4 liters of thick, hazy orange-red, peritoneal fluid. Subsequent paracentesis only yielded 700 mL of fluid or less each time. Fluid studies revealed SAAG 3.4, Total Protein 0.4. Indicating cirrhosis as the initial cause of the ascites. Several imaging studies including ultrasound, CT, MRI of the abdomen and pelvis revealed complex, loculated, septated ascites versus cystic masses of peritoneal/ovarian origin. Complicating the diagnosis was that the initial cytology was negative. However, not believing that this was simply cirrhosis, we sent the subsequent paracentesis for cytology, and the repeat cytology demonstrated abundant mucin and mucin balls, concerning for gastrointestinal or gynecologic neoplasm. Endoscopy by Gastroenterology revealed bulging of the appendiceal orifice, but no discrete masses were visualized. She was evaluated by Surg-Onc, Gyn-Onc, and Urology and underwent exploratory laparotomy with resection of an intraabdominal mass >10 cm, with diffuse mucin through the abdominal cavity. She received intra-operative chemotherapy/HIPEC with mitomycin C after debulking therapy and ureteral stents. Final pathology revealed low grade appendiceal mucinous neoplasm.

Discussion: This case illustrates the importance in trusting your clinical judgement when evaluating all of the data collected and the importance of a multidisciplinary approach to complicated cases. Based on the SAAG and total protein of the fluid, the diagnosis was cirrhotic ascites. However, the appearance of the fluid removed was not consistent with that diagnosis. By trusting our clinical judgement and discussing the case with multiple disciplines we were able to come to the final diagnosis and improve the quality of life of our patient. Pseudomyxoma peritonei is a rare condition, with increasing abdominal girth as the most common presenting symptom, and ultimately requiring surgical debulking with intraperitoneal chemotherapy for diagnosis and treatment.
A Case of Antisynthetase Syndrome Initially Presenting as Recurrent Pneumonia

Authors: Jennifer Kennard, DO and Shweta Kishore, MD

Introduction: Antisynthetase syndrome is a rare immune-mediated disorder that is seen in patients with myositis who have antibodies against aminoacyl-tRNA synthetases. Here we present a case of a patient who was treated for recurrent multifocal pneumonia but was later diagnosed with antisynthetase syndrome.

Case Presentation: A 41-year-old female presented to the emergency department after a fall. She reported that her legs “gave out” causing her to fall to the ground. In addition, she reported increasing shortness of breath and cough over the past several weeks. Prior to this presentation, she had been admitted to the hospital with dyspnea, cough and weakness twice in the past four months. During the two previous admissions, imaging was consistent with multifocal pneumonia. She was treated with antibiotics and steroids but would again feel worse after completing treatment.

Upon this presentation, physical exam was significant for hypoxia, coarse rales over bilateral lung bases and 3/5 muscle strength in the proximal muscles of the upper and lower extremities. Dermatologic exam was benign. Computed tomography of the chest revealed pulmonary emboli and diffuse bilateral ground glass opacities and focal consolidations. She was started on anticoagulation and broad-spectrum antibiotics while further work-up was initiated to investigate the cause of recurrent pneumonia in an otherwise healthy adult. Bronchoalveolar lavage did not reveal any organism. Human immunodeficiency virus, mycobacterium tuberculosis, and fungal antigens returned negative. Anti-nuclear antigen (ANA) was negative. Extractable nuclear antigen panel returned positive for anti-Jo-1 antibody. Creatinine kinase and aldolase levels were elevated at 3,450 and 106.4 U/L respectively. Magnetic resonance of the femur revealed mild muscular edema and enhancement of the muscles of the pelvis and femurs. Muscle biopsy revealed inflammatory infiltrates extensively throughout the specimen consistent with polymyositis. She was started on treatment for interstitial lung disease (ILD) and inflammatory myositis with high dose steroids, intravenous immunoglobulin and mycophenolate mofetil. The patient quickly responded to treatment with improvement in dyspnea and muscle weakness. Creatinine kinase and aldolase improved to 170 and 5.9 U/L respectively at follow-up four weeks later.

Discussion: Features of antisynthetase syndrome can include myositis, ILD, symmetric non-erosive arthritis, and mechanic’s hands. Patients will often have a negative ANA as the antisynthetase antibodies stain cytoplasmic antigens. Of these antibodies, anti-Jo-1 is the most common. Myositis and ILD are the most common presenting features of antisynthetase syndrome and in up to 20% of cases ILD may be the initial presentation as seen in this case. This patient’s inflammatory myositis quickly progressed causing her significant muscle weakness at the presentation described here; however, her initial symptoms were dyspnea and cough. In a patient presenting with dyspnea and recurrent pneumonia not responding to antibiotics, antisynthetase syndrome should be on the list of differential diagnoses.
When the Bottom Falls Out: Acquired Hemophagocytic Lymphohistiocytosis in the setting of Congenital Panhypopituitarism

Authors: Ryan P. Collier, MD; Joshua Tunnage, MD; Ian Ward, MD; Matthew Wright, MD

Introduction: Acquired Hemophagocytic Lymphohistiocytosis (aHLH) is a rare condition that has a significantly high mortality despite prompt treatment with high dose steroids and in some cases requires chemotherapy to downregulate an overactive immune system causing excessive inflammation and tissue destruction by activated macrophages and lymphocytes.

Case Presentation: Fifty-seven year old caucasian female with a history of panhypopituitarism, previously on hydrocortisone that was stopped several months prior to presentation, and cutaneous vasculitis treated with dapsone and colchicine, presented to the emergency room complaining of fevers, cough, and malaise. Due to concern for sepsis secondary to community acquired pneumonia, broad-spectrum antibiotics and stress dose hydrocortisone were initiated. Patient symptomatically improved during the first twenty four hours, however developed oliguria and two distinct maculopapular rashes, one across both forearms which blanched and one nonblanching rash across the chest which spread to the abdomen and legs. Dermatology and Rheumatology were consulted, with the arm rash consistent with previous cutaneous vasculitis and truncal rash thought to be a vancomycin drug reaction. Initial chest radiograph showed potential pneumonia with subsequent CT Chest/Abdomen/Pelvis showing no acute pulmonary process but potential cholecystitis. Vancomycin was discontinued but patient remained on piperacillin-tazobactam. Right upper quadrant ultrasound revealed potentially biliary sludge and features concerning for acalculous cholecystitis.

Patient became anuric and continued to clinically deteriorate. Abdominal rash continued to worsen, patient developed bicytopenia, hyperferritinemia, and DIC like physiology eventually requiring blood product transfusions. Due to clinical concern for possible acquired HLH and an Hscore of 210 indicating a 93% probability of having acquired HLH, the patient was started on 1 Gram of intravenous methylprednisolone daily. Laboratory analysis which supported this diagnosis consisted of ferritin >7500 ng/mL, LDH 2089, hypofibrinigenemia, and bicitiopenia. Soluble IL-2 Receptor alpha was 32,768 U/mL (223-710) with values greater than 10,000 U/mL having a 93% specificity to rule in HLH. Hematology was consulted but bone marrow biopsy unable to be performed due to patient’s clinical status. Patient’s acidosis continued to worsen requiring intubation and CRRT dialysis. Despite escalation of intensive care, cardiac arrest occurred with eventual patient demise.

Discussion: Acquired hemophagocytic lymphohistiocytosis is a life threatening disease that can mask as sepsis and can be difficult for providers to identify and treat in a timely manner. Patient’s case was complicated by known prior cutaneous vasculitis at baseline, with new rash thought to be a drug reaction, thus delaying high dose steroid administration. This disease proves difficult to diagnose and has a high mortality despite aggressive treatment. The Hscore can be an excellent tool which can lead to earlier recognition and treatment in the disease course which may potentially lead to improved outcomes. In the end, it is imperative that aHLH be maintained in the differential diagnosis of immunocompromised patients presenting with sepsis like physiology.

References

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A case of primary genital herpes as the nidus for HSV hepatitis and hemophagocytic lymphohistiocytosis

Authors: R. Clark Cutrer, MD; W. Fowler Joiner, MD; R. Noelle Bates, MD; Shradha Ahuja, MD

Introduction: Primary genital herpes is a common manifestation of Herpes Simplex Virus (HSV), typically HSV-1 or 2. In rare instances, primary genital herpes can develop into disseminated HSV infection, especially in immunocompromised patients. Disseminated HSV can manifest itself in many forms, including hepatitis and can even cause Hemophagocytic Lymphohistiocytosis (HLH). Here we present a case of primary genital herpes leading to HSV hepatitis and ultimately HLH in an immunocompromised patient with lupus.

Case Presentation: Patient is a 25yo African-American female with a history of Juvenile Rheumatoid Arthritis and Systemic Lupus Erythromatous (SLE) on chronic immunosuppression who presented as a direct admission from clinic on 4/13 for fever of unknown origin. Patient revealed that she had noticed a singular genital ulcer. OB/Gyn was consulted, and by exam, clinically diagnosed patient with HSV. Patient was started on PO acyclovir (400 mg TID) initially, but was soon escalated to 575 mg IV TID. Vaginal culture came back positive for HSV-2. On Hospital Day 2, liver enzymes dramatically increased into the 1000s, concerning for a viral hepatitis. CT chest w/ abdomen w/ pelvis was obtained on 4/14, which was most significant for miliary lesions in liver. The clinical picture was thought to be possibly 2/2 disseminated HSV, so Infectious Disease was consulted on 4/16. Infectious work-up was undertaken. HSV PCR was ultimately positive for HSV type 2 (2x10E7 copies). On 4/20, patient underwent liver biopsy, which demonstrated necrosis of liver parenchyma with consistent with viral hepatitis; HSV I & II were positive in the biopsy. Per ID recommendations, patient completed 21 days of IV Acyclovir followed by PO Valacyclovir for prolonged course. She went on to develop HLH in the setting of disseminated HSV infection, which was proven by bone marrow biopsy, and subsequently resolved with treatment of underlying HSV.

Discussion: HSV is a very rare but serious complication of primary HSV infection seen predominantly in immunocompromised patients with a mortality rate up to 80-90%. Early clinical recognition is of utmost importance because early treatment with Acyclovir has shown in an improvement in mortality and in progression of liver disease. Another rare clinical entity demonstrated in this case is disseminated HSV as the precipitating cause of HLH. Hemophagocytic lymphohistiocytosis is a rare but very serious clinical condition that typically arises in adults as a result of an underlying infection, malignancy or autoimmune process. Without proper treatment, HLH carries a very high mortality, up to 70%. In our patient, treating the HLH meant treating the underlying infection, which proved successful with full resolution.
MISSISSIPPI CLINICAL VIGNETTE POSTER FINALIST - PRIYA PATEL, MD

Perhaps it’s time to become vegetarian: Alpha-gal mediated red meat allergy

Authors: Priya Patel M.D., Clark Cutrer M.D., Jackson Ross M.D., Taylor Harvey, Patricia Stewart M.D., Amanda Clark M.D., Department of Medicine at University of Mississippi Medical Center and School of Medicine at UMMC

Introduction: A rare, IgE-mediated allergy to galactose-alpha-1,3-galactose (“alpha-gal”) is thought to develop after a tick bite and result in a new, adult-onset red meat allergy. Alpha-gal anaphylaxis can be challenging to diagnose and can be easily overlooked due to the delayed onset of symptoms three to six hours after exposure, unlike most IgE-mediated reactions.

Case Presentation: A 66-year-old male with Gliobastoma presented to the Emergency Department (ED) after experiencing flushing, pruritus, hives, mild dyspnea, lightheadedness and hypotension as measured by his home blood pressure machine. He had no known allergies and denied any new exposures except for several tick bites in the past with the most recent bite about two months ago and initiation of two chemotherapy agents, Temozolomide and Lomustine, about three months prior. Earlier in the day, he had a hamburger steak and mashed potatoes. Additional history revealed that about 2 months ago, he had a similar previous episode that was less severe with hives, intense pruritus, nausea and vomiting several hours after eating a sausage biscuit. Physical exam revealed generalized erythema of trunk and extremities, lungs clear to auscultation bilaterally and no facial or oropharyngeal edema. He was treated with IV diphenhydramine and prednisone and did not require epinephrine due to normalized vital signs. Temozolomide was initially considered as the culprit; however, the timeline for development of symptoms did not fit that of a classic IgE-mediated reaction. He was admitted overnight for observation and remained asymptomatic. At discharge, patient was prescribed an epinephrine autoinjector in case of recurrence and referred to allergy clinic. Lab tests in clinic showed elevated alpha-gal IgE >100 kU/L, pork IgE 19.8 kU/L (Class 4-Strongly Positive) and beef IgE 43.4 (Class 4-Strongly Positive). Based on the timeline, it was felt that he likely had an anaphylactic reaction to the red meat after sensitization to galactose-alpha-1, 3-galactose (alpha gal) from a tick bite. He was informed to avoid red/mammalian meat with further plans to add fexofenadine and famotidine if symptoms recurred.

Discussion: Anaphylaxis is a life-threatening hypersensitivity reaction with incidence of 49.8 cases per 100,000 person years in the United States. The two most important tenants in management of anaphylaxis are prompt treatment to alleviate symptoms and the avoidance of triggers. The characteristics of red meat allergy are different from typical allergic reactions in that patients do not develop symptoms until 3-6 hours after the ingestion. Despite this difference from the usual Ig-E mediated reaction, symptoms can be severe or even life-threatening. This case illustrates the significance of considering new-onset of red meat allergy even in adults who have been exposed to red meats prior and especially in patients with an unclear cause for anaphylaxis.

References

Hemophagocytic Lymphohistiocytosis: A Severe Clinical Presentation after a Tick Bite

Authors: Zarir Ahmed, DO., Rachna Rawal, MD., Martin Schoen, MD., Kevin Palka, MD.

Introduction: Hemophagocytic Lymphohistiocytosis (HLH) is an uncontrolled inflammatory response associated with a wide variety of conditions sometimes including infection. However there have only been a few case reports where tick bites cause HLH. This case describes a patient bitten by a tick and develops HLH and candidemia.

Case Presentation: A 40 year-old male went camping in southeast Missouri in May and suffered tick bites. He developed a fever, rash and myalgias. He was treated with doxycycline, which was discontinued due to anaphylaxis. Due to worsening abdominal pain and fatigue he went to the hospital. He was febrile, ill-appearing, and jaundiced. He had scattered petechiae bilaterally on his axillae and flanks. His abdominal exam was unremarkable. Laboratory evaluations revealed significant hyperbilirubinemia, elevated liver enzymes, thrombocytopenia, and neutropenia. He was empirically started on ceftriaxone for a tick-borne illness. An abdominal CT revealed hepatosplenomegaly and ascites. Infectious evaluation for viral hepatitis and tick-borne illnesses was negative. Due to clinical deterioration, his antibiotics were broadened and treatment for idiopathic thrombocytopenia purpura with intravenous immunoglobulin and steroids was initiated. However, the patient developed shock and expired after cardiac arrest.

Autopsy revealed multi-organ fungal growing Candida tropicalis. Pathology suggested hemophagocytic lymphohistiocytosis (HLH) in the liver, spleen, lymph nodes and bone marrow. He met 6/8 criteria for HLH with fever, splenomegaly, thrombocytopenia and neutropenia, elevated ferritin, elevated interleukin-2 receptor, and low natural killer cell activity.

Discussion: The diagnosis of HLH must fulfill five out of nine diagnostic criteria including fever, splenomegaly, bi- cytopenia, hypertriglyceridemia, hypofibrinogenemia, elevated ferritin, hemophagocytosis in the bone marrow/spleen/lymph nodes, low natural-killer cell activity, and elevated interleukin-2 receptor. Treatment of HLH involves immune-suppression with chemotherapy to suppress the hyperinflammatory and supportive care such as transfusions.

The immediate cause of death in our patient was widespread fungal infection secondary to his profound pancytopenia and liver failure. The etiology of his pancytopenia is likely related to a tick-borne illness which led to the development of HLH.

There have only been a few case reports from tick-borne related illness causing HLH in areas such as Southeast Asia and the Midwest United States. This case serves as a reminder that HLH can present as a severe clinical presentation from tick-borne illnesses. This likely predisposed the patient to develop disseminated Candida. This case emphasizes the importance to recognize the diagnostic criteria for HLH. In addition, in patient with severe neutropenia, it is important to consider beginning antifungal medications for prophylaxis. Early diagnosis, treatment and supportive care for HLH and co-infections can improve prognosis.
A Diagnostic Terror... A Teratoma Producing Anti-NMDA-R Antibodies

Authors: Neil Evans MD1, Amitabh Singh, DO1, Cory Cheatham MD1, 1Saint Louis University, Saint Louis, MO

Introduction: Autoimmune phenomena in the brain are rare and often a diagnostic challenge. We present a case of Anti-N-methyl-d-aspartate (NMDA) receptor encephalitis that illustrates this challenge.

Case Presentation: A 23 year old female with an unremarkable past medical history presents to our hospital with psychosis. She was recently discharged from a local psychiatric facility for unresolved “Flakka” intoxication.

On presentation she was encephalopathic, aggressive, and inappropriate to staff. Initial workup including EEG was unrevealing. MRI of the brain was negative for malignancies and other abnormalities. LP was performed but was negative for common bacterial and viral infectious causes. CT of the abdomen and pelvis was performed demonstrating a right sided ovarian cyst. Pelvic MRI was performed to better characterize the cyst and it had characteristics consistent with a teratoma. A repeat LP demonstrated an elevated oligoclonal bands and 1:20 Anti-NMDA-antibody titre. A presumptive diagnosis of NMDA-R encephalitis was made. She was treated with plasmapheresis, IV immunoglobulin and IV solumedrol. She underwent oophorectomy; four days after she began showing signs of improvement in her cognition and by two weeks afterwards, the patient had returned to her baseline mental status and was discharged home.

Discussion: Anti-NMDA-receptor encephalitis is the leading reversible cause of encephalitis in females under the age of 30. MRI is not sensitive for this condition with 50% appearing normal. Diagnosis is made by lumbar puncture looking for the antibody. Most cases of Anti-NMDA-receptor encephalitis have an identifiable ovarian teratoma, which are thought to contribute to an immune-mediated pathogenesis. Treatment protocols emphasize supportive care and teratoma removal. The treatment regimen for anti-NMDA-receptor encephalitis includes high dose IV glucocorticoids, intravenous immune globulin and plasma exchange. In our case the patient received appropriate therapy and had an identifiable teratoma which was excised; leading to her positive outcome. It is important that internists consider anti-NMDA receptor encephalitis in their evaluation of young females with unknown encephalitis as once corrected the prognosis is good.

References

Acute Upper Gastrointestinal Bleeding: An Atypical and Potentially Life-Threatening Presentation of Granulomatosis with Polyangiitis

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Introduction: Granulomatosis with polyangiitis (GPA)—previously known as Wegener granulomatosis—is a rare autoimmune disorder of an unknown etiology [1]. The main findings are necrotizing granulomatous inflammation and ANCA vasculitis in small and medium-sized blood vessels [1]. The disease typically presents with respiratory and renal manifestations; however, GPA may also rarely present with GI bleeding [1] [2] [3] [4].

Case Presentation: A 48-year-old Caucasian man with a 30 pack-year smoking history and COPD presented to the hospital with complaints of worsening dyspnea and generalized abdominal pain. He also reported nausea, weight-loss, and a rash on his upper extremities. He denied any significant alcohol or NSAID use. He was tachycardic upon admission and slightly hypotensive. The patient’s physical exam was notable for bibasilar lung crackles, diffuse abdominal tenderness, and purpuric lesions on his hands and arms. His laboratory work-up revealed acute kidney injury, leukocytosis, anemia, an elevated CRP and ESR. The patient was started on aggressive intravenous fluids, blood transfusions, and a proton pump inhibitor. A subsequent EGD revealed a bleeding gastric ulcer and was treated endoscopically. CT chest and abdominal imaging revealed numerous bilateral pulmonary nodules, a right cavity lung lesion measuring 2.6 x 2.1 cm, calcified mediastinal and hilar lymph nodes. Considering the patient’s presentation and work-up, a vasculitis investigation was initiated. The patient’s proteinase 3 antibody (PR3-ANCA/c-ANCA) value was elevated. The patient was started on pulse methylprednisolone for a suspected GPA. The patient underwent a right lung nodule biopsy which was determined to be necrotic and non-diagnostic. A left kidney biopsy was obtained and pathologically determined the presence of pauci-immune complex trapping and focal necrotizing glomerulonephritis with crescent formation. The diagnosis of GPA was made and the patient’s symptoms improved with pulse methylprednisolone. The patient was later discharged with close outpatient follow-up.

Discussion: GPA is a rare condition that is typically associated with renal and pulmonary complications. Acute upper gastrointestinal bleeding is possibly a life-threatening presentation of patients with GPA that requires prompt diagnosis and intervention [1]. As presented in this case report, a patient’s plan of care should focus upon hemodynamic stability, possible gastrointestinal endoscopic evaluation and repair, pulse steroids, and biopsy confirmation.

References

MISSOURI CLINICAL VIGNETTE POSTER FINALIST - PUJA JADAV, MD

When Statins Attack: A Case of Necrotizing Autoimmune Myopathy

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Introduction: Statins are commonly prescribed medications that reduce cardiovascular disease. Their side effect profile consists of muscle-related adverse effects that are classified into toxic or autoimmune forms. We present a case of statin-induced necrotizing autoimmune myopathy (SINAM), a recently characterized entity within this spectrum of statin myotoxicity. While rare, it is important to recognize SINAM as a potential side effect of statin use and identify the main constellation of findings that characterize it.

Case Presentation: A 58-year-old African American man with a history of cervical spondylosis, diabetes, hyperlipidemia presented with lower extremity swelling and progressive leg weakness following a fall one month prior whereby his legs “gave out”. He had been restarted on atorvastatin 1 year prior, which had initially been discontinued for muscle spasms then deemed unrelated. Physical exam was significant for 3+ edema of lower extremities bilaterally to thighs and proximal muscle weakness with strengths of 4/5 in biceps/triceps but 3/5 in hip flexors/extends. Pertinent workup for his weakness revealed a benign endocrine workup, normal inflammatory markers (ESR 12, CRP 0.26), negative myositis panel, but a positive ANA, greatly elevated muscle markers (Aldolase 66, CK 11,635), and a high titer positivity for Anti-HMG CoA Reductase (>200). MRI femur showed increased signal within proximal posterior and medial muscle groups indicative of compartmental myositis and skeletal muscle biopsies revealed a necrotizing myopathy with a paucity of chronic inflammation. The patient was started on prednisone 50mg daily, all statins added as allergies, and was discharged with close rheumatology follow up. Given his continued falls despite intense physical therapy, mycophenolate and IVIG were added, with improvement in strength and edema, a downtrend of his CK to 400, and the ability to taper off prednisone.

Discussion: The pathogenesis of SINAM is via the marked up-regulation of the hydroxy-3-methylglutaryl-coenzyme A reductase receptor protein (HMGCR) in regenerating muscle cells of susceptible persons when exposed to statins, which then act as autoantigens to induce antibody production. The anti-HMGCR antibody then triggers necrosis and regeneration of muscle fibers, resulting in severe muscle damage that persists even after the discontinuation of the statin. SINAM is characterized by significant proximal muscle weakness, CK levels over 6000 U/L, evidence of muscle-cell necrosis on biopsy with minimal inflammatory infiltrates, and the presence of autoantibodies against HMG-CoA reductase as demonstrated in our patient case. Treatment includes discontinuing the statin, with half of all patients needing triple therapy consisting of high dose prednisone and two immunosuppressive agents. When recognized and treated, most patients have good outcomes. While inflammatory myopathies are rare, with SINAM having an incidence of 2 out of 100,000 persons on statins, given the large prevalence of statin use, the important of early recognition and appropriate management of this debilitating condition cannot be under emphasized.

References

HIV-Associated Vacuolar Myelopathy and HIV-Associated Dementia as the Initial Manifestation of HIV/AIDS

Authors: Natalia Wuliji, DO; Jason M. Lunt, DO; Matthew J. Mandell, DO; Abigail L. Carlson, MD

Introduction: HIV-associated vacuolar myelopathy (HIV-VM) is the most common cause of spinal disease in HIV/AIDS. HIV-VM causes progressive spastic paraparesis, sensory ataxia, and autonomic dysfunction. It is a progressive myelopathy that shares features with subacute combined degeneration, seen in vitamin B12 deficiency as well as other neurological diseases and can occur synchronously with HIV-associated dementia (HAD). Here, we describe a rare case in which a patient’s initial presentation of HIV/AIDS was both HIV-VM and HAD.

Case Presentation: A fifty-three-year-old man presented with a six-month history of numerous falls due to progressive gait instability with associated memory loss, tremor, urinary retention, and impotence. His exam revealed hyperreflexia and weakness in bilateral lower extremities, positive Babinski sign, dysmetria, and ataxic gait. Imaging was notable for an MRI-brain that demonstrated non-specific volume loss and diffusely increased T2 signal throughout the supratentorial white matter. A cervical spine MRI demonstrated degenerative disease and moderate central canal stenosis; however, patient had no neck, arm, or radicular pain, and Lhermitte’s sign was absent – making these findings unlikely to explain his profound symptoms. Lumbar puncture showed isolated lymphocytic pleocytosis with all other CSF testing unremarkable. Thyroid and vitamin B12 testing were unrevealing, as was an expansive infectious etiological evaluation including syphilis testing, Lyme disease serologies, and endemic fungi serologies. Heavy metal testing was also unremarkable. His HIV testing ultimately returned positive. He was diagnosed with HIV-VM and HAD which improved after several months of anti-retroviral therapy.

Discussion: HIV-VM and HAD presenting as the first sign of HIV/AIDS is uncommon. HIV-VM and HAD frequently progress in parallel. Similar pathophysiologic mechanisms may underlie HIV-VM and HAD. HAD has been characterized as a subcortical dementia, leading to cognitive, behavioral, and motor dysfunction. Initial symptoms may be subtle and are often overlooked or misdiagnosed as depression. Motor dysfunction may manifest early on as frequent stumbling and tripping. While HIV-VM can present at any stage of HIV infection, it is most commonly seen in advanced disease. The diagnosis of HIV-VM is a diagnosis of exclusion, therefore requiring an evaluation for many other etiologies, which was performed in this case. Interestingly, there are some MRI findings that are associated with the diagnosis which include spinal cord atrophy increased signal on T2-weighted images in the white matter usually affecting the dorsal columns and lateral corticospinal tracts of the cervical and thoracic cord. A normal appearing spinal cord does not, however, exclude the diagnosis. While it is routine to evaluate patients with a sensory neuropathy for thyroid disorders, diabetes, and vitamin B12 deficiency, evaluating for HIV in this population is less commonly pursued. This case highlights the importance of considering HIV testing in a patient with a sensory neuropathy and/or progressive cognitive impairment.
Is Eculizumab the Next Clue in Treating Typical Hemolytic Uremic Syndrome?

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Introduction: Typical Hemolytic Uremic Syndrome (HUS) is caused by Shiga Toxin-producing E. Coli whereas atypical HUS is complement mediated. Eculizumab is a monoclonal antibody to complement factor C5 that blocks complement activation and its use in atypical HUS has been well established. However, its use in STEC-HUS remains controversial and at times as a last resort. The following case highlights the use of Eculizumab in a patient with STEC-HUS with severe neurological symptoms.

Case Presentation: An 18-year-old prior healthy female developed foul-smelling flatus approximately 5 days prior to a trip to the Dominican Republic. On the morning of her departure she developed loose stools and felt weak. She had a syncopal episode while waiting in line to board the plane in Charlotte, North Carolina. Subsequently, she traveled to the Dominican Republic where she developed bloody diarrhea within two hours of arrival. She was seen at the local ER where she received IV hydration, Ciprofloxacin, and Flagyl. She was transferred to the ICU because of hypotension and tachycardia and had persistent diarrhea along with abdominal pain. Arrangements were made to transfer the patient to St. Luke’s Hospital because of her acute kidney failure and difficulty in managing her care. Prior to transfer, she had a seizure. She was intubated and treated with Keppra. She was life-flighted to St. Luke’s Hospital.

Upon arrival labs revealed a leukocytosis of 30.7 K/uL, hemoglobin of 9.0 g/dL, platelets of 68 K/uL, BUN 55 mg/dL, creatinine 5.1 mg/dL, LDH 9606 U/L, haptoglobin < 20 mg/dL, fibrinogen 515 mg/dL. Enteric Pathogen PCR panel was positive for Shiga Toxin 2, and a diagnosis of Hemolytic Uremic Syndrome was made. Antibiotics were stopped, and treatment with Eculizumab and hemodialysis was started. She remained unresponsive even off sedation for 72 hours. An EEG showed an encephalopathic pattern. Follow up brain MRI showed diffuse hypoxic-ischemic injury, including diffuse areas of diffusion restriction involving the thalami, splenium, pons, and brainstem. Her neurological function improved slowly, and her creatinine and hemoglobin returned close to the baseline.

On the 12th ICU day, she was extubated and transferred to the medical floor where she continued to recover. On day 19 of her hospitalization the patient was discharged home. After discharge, she continued outpatient hemodialysis. Subsequently, her creatinine returned within normal range and dialysis was discontinued and the patient made a full recovery.

Discussion: This patient had STEC-HUS with severe CNS involvement. Several meta analyses have shown that in patients with typical HUS and CNS involvement early use of Eculizumab appears to improve neurological outcome. The benefits of treating this young previously healthy patient with Eculizumab outweighed the risks. Given the recent outbreak of STEC and HUS, further studies focusing on the use of Eculizumab in STEC-HUS are warranted.
A Case of Unexplained Coagulopathy

Authors: Amitabh Singh, DO1 Neil Evans MD1, Jennifer Schmidt MD1, 1 Saint Louis University, Saint Louis, MO

Introduction: Use of synthetic illicit drugs, including marijuana, is on the rise. Commonly these drugs are laced with additives, many of which can have severe hematologic effects, to enhance the drugs’ euphoric effects. Here we describe a case of spontaneous hemorrhage after ingestion of marijuana laced with brodifacoum.

Case Presentation: A 25-year-old male presented with bilateral lower quadrant abdominal pain radiating to his back starting one day prior to presentation. He also reported hematuria, decreased appetite. He denied new foods, medications, supplements or illicit substances and travel.

On presentation, vital signs were normal. Abdomen was distended and tender. Computer tomography showed bilateral acute perinephric hemorrhage extending into the retroperitoneum and pelvis. Lab work showed Hgb:7.1 platelets 178, PTT >212, PT >106, INR >10. Urine drug screen was negative.

The patient was admitted to the ICU and given fresh frozen plasma (FFP) and IV vitamin K. Workup showed factors II, IX, and X below 3 (normal range 65-175) and normal levels of factors V and VIII. Warfarin toxicity panel was positive for brodifacoum. PTT and INR trended down and normalized. Patient was discharged with oral vitamin K.

On further discussion, the patient admitted to smoking synthetic marijuana.

Discussion: As synthetic marijuana is not detected on standard urine drug screens, its use is increasing. Often, substances are added to increase or prolong its euphoric state. One such additive is brodifacoum, a coumadin-derived rodenticide. It inhibits the enzyme vitamin-K-epoxide reductase (VKOR), as mediator in the production of factor II, VII, IX, and X, resulting in low factor levels. Toxicity typically occurs by ingestion of laced-marijuana and symptoms directly correlate with amount ingested. Treatment is focused on repletion of Vitamin K with the addition of FFP for severe toxicity. During 2018, there have been publicized outbreaks of laced marijuana resulting in multiple deaths. It is critical that internists are able to identify when patients present after ingestion of laced-illicit drugs—not only does this allow for appropriate workup and treatment, it is also essential in identifying public health emergencies.

References

A Rare Etiology of Hypercalcemia

Authors: Kimberly Ng, MD, Alyssa Burkhart, MD, FACP, Nolawit Tesfaye, MD

Introduction: Common causes of hypercalcemia include hyperparathyroidism, malignancy, and granulomatous disease. Though infrequently reported, the osteolytic effect of vitamin A on bone can also cause hypercalcemia if ingested in excessive quantities.

Case Presentation: We present a unique case of a 52-year old woman who presented to the Endocrinology clinic following a referral from her primary care provider for intermittent hypercalcemia and persistently low parathyroid hormone (PTH). Symptoms on her initial visit included chronic fatigue, constipation, and a 10-pound weight gain over the last year. Past medical history was significant for nephrolithiasis, renal colic, toe fractures, and depression. Supplements included a daily multivitamin and vitamin D3 5000 IU. In addition to consuming meals from a pre-prepared meal service, her diet consisted of 2 glasses of milk per day plus another serving of milk with cereal. Family history was negative for hypercalcemia. Physical examination was unremarkable. As part of her initial workup, calcium and PTH levels were re-drawn. As previously observed, these lab values were mildly high and low, respectively. However, her ionized calcium level was within the high-normal range. Despite this result, a complete workup for hypercalcemia was pursued due to her lengthy 9-year history of abnormal lab findings. PTHrP, vitamin D metabolites, SPEP, and serum-free light chain assay all returned normal. In contrast, her vitamin A level was notably elevated. After further discussion with a registered dietitian, it was determined that her meals varied between 225 IU to 5436 IU of vitamin A content (recommended daily allowance for vitamin A for a female aged over 50 years is 2333 IU). Her multivitamin contained an additional 3500 IU of vitamin A. Based on these findings, it was concluded that she was consuming excessive quantities of vitamin A. She was advised to discontinue her multivitamin and avoid liver, fish, eggs, and fortified milk and cereal. While her symptoms of fatigue persisted at her follow up appointment 2 months later, her calcium levels returned to normal and vitamin A levels dropped. The patient’s clinical signs and pertinent labs are continuing to be monitored.

Discussion: This clinical vignette describes a rare case of hypercalcemia secondary to hypervitaminosis A. It highlights the value of a comprehensive workup that includes vitamin A levels when routine lab testing fails to identify the etiology of hypercalcemia.
Acyclovir for Zoster: Friend or Foe?

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Introduction: Determining the etiology of altered mental status in patients with end-stage renal disease and Zoster meningitis can be difficult. In complex clinical cases, it’s important to consider a broad list of differential diagnoses, including potential uncommon medication adverse effects. Bold clinical decision-making in ambiguous diagnoses can be life-changing and result in better patient outcomes.

Case Presentation: A 50-year-old man with history of end-stage renal disease developed several days of increasing somnolence. His prolonged hospitalization had been complicated by profound gastrointestinal bleed, hypercapnic respiratory failure, and ischemic hepatitis. Additionally, he developed Janeway lesions and a mitral valve vegetation with negative blood cultures, prompting empiric treatment for endocarditis with vancomycin (15mg/kg dosed according to levels) and cefepime (1g with dialysis). Two days later, the patient was diagnosed with Zoster and started on renally-dosed valacyclovir (500mg daily).

Within several days of antimicrobial initiation, the patient became more lethargic and developed myoclonus. Given clinical Zoster rash, lumbar puncture was obtained to evaluate for encephalitis. His lethargy and myoclonus improved following each round of dialysis, raising concern for cefepime neurotoxicity, and cefepime was discontinued (transitioned to ceftriaxone). Meanwhile, the patient’s cerebrospinal fluid Zoster PCR was positive, and he was started on IV acyclovir (5mg/kg/24h). Over the next several days following cefepime cessation, the patient’s myoclonus resolved, and mental status returned to baseline, with the most notable improvements following dialysis.

These improvements were short-lived, and the patient soon became increasingly somnolent again, declining to the point where he was no longer responsive to painful stimuli. Subclinical seizures were excluded; potential CNS depressant medications (including gabapentin) were discontinued, but his mental status failed to improve. Acyclovir neurotoxicity was considered, and on treatment day 8, given no other plausible alternatives, acyclovir was discontinued. By the following day, he had already improved. After two dialysis runs, he was alert, oriented and back to baseline.

Stored serum obtained before acyclovir discontinuation was analyzed by two separate laboratories for 9-carboxymethoxymethylguanine (CMMG) level, an acyclovir metabolite. Both laboratories reported abnormally high levels of CMMG, consistent with severe accumulation and increased risk of acyclovir neurotoxicity.

Discussion: Two aspects of this case make it especially unique. First, this case reflects the diagnostic dilemma determining whether the patient’s symptoms are related to the condition, Zoster meningitis, or the treatment of the condition, IV acyclovir. Inadequately treated zoster meningitis can be devastating. However, persistent treatment resulting in neurotoxicity is also harmful. Second, this case raises the question; does development of neurotoxicity with one drug predispose to neurotoxicity from other drugs? Altered mental status is a condition seen frequently by the general internist. Neurotoxicity comprises only a fraction of those cases; thus, a broad differential must be considered when assessing for possible drug-induced neurotoxicity.
Worth Waiting for a Diagnosis

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Introduction: Pulmonic tularemia is a rare and severe infection with mortality rates of 30-60% due to acute respiratory failure. False negative results occur in the acute phase, therefore titers are not reliably positive until two weeks of active infection.

Case Presentation: A 29 year-old man presented with a four day history of vomiting, diffuse abdominal pain, and confusion. His past medical history was unremarkable. Social history included occupation in landscaping and recreational use of marijuana and alcohol. On exam, he was febrile at 39.6°C and tachycardic. He had an inflamed right molar. Neurologic exam revealed distraction and confusion but no focal deficits. Labs were significant for leukocytosis with 52% bands, sodium 124, creatinine 1.82, AST 2697, ALT 471, CK >80,000, procalcitonin 41.22, and urinalysis suggesting myoglobinuria. Chest CT revealed multifocal consolidation throughout the lungs. He deteriorated quickly and was admitted to the ICU for aggressive fluid resuscitation. Empiric antibiotic treatment was initiated with IV vancomycin and piperacillin-tazobactam.

Infectious disease was consulted and extensive microbiologic assays were ordered. Piperacillin-tazobactam was discontinued while gentamicin, doxycycline, levofloxacin, and ceftriaxone were started. Daily procalcitonins peaked at 92.76. His temperature maxed at 40.1°C. Over the next two days, he became increasingly encephalopathic with agitation, dysarthria, and lack of orientation. All diagnostic studies were negative.

After five days of ICU care, his mentation improved and was transferred to the floor. He had residual ataxia and dysarthria. He continued to improve over the next week. After eleven days in the hospital, the tularemia panel was repeated. Repeat imaging revealed persistent lung opacities and new pleural effusion. His treatment was reduced to monotherapy with doxycycline. The repeat tularemia panel came back positive on day 16 and he was discharged on day 17. The remaining 5 days of a 21-day course of doxycycline was prescribed at discharge.

Discussion: Tularemia manifests as six different subtypes, which physicians need to be aware of for purposes of early diagnosis and treatment. This patient was diagnosed with the pulmonic form of tularemia, which has a high mortality rate. His diagnosis was confirmed after more than 20 days of symptoms, due to the prevalence of false negatives in the acute phase. He presented with rhabdomyolysis and encephalopathy, rapid deterioration, and late pulmonary symptoms. Without early treatment with gentamicin, this patient would have likely had adverse outcomes. The most likely epidemiologic explanation for this case is contact with infected rodents while landscaping.

There were 230 cases of Tularemia reported in the U.S in 2016. The incidence over the last 10 years is 0.7 per 100,000 in the U.S. Overall mortality rate for untreated tularemia is <8%. It decreases to <1% with antibiotic treatment. Early empiric antibiotic treatment and proper diagnosis is key when dealing with this specific zoonotic diseases.

References

Autoimmune Hepatitis in an Immunocompromised Patient

Authors: Annie S. Hong, Jenny M. Hong, Wen Yuan Yu, Yen Cao, Carina Chang, Ahl Jeffrey Caseja, Awad Javaid, Badrunissa Hanif

Introduction: Autoimmune hepatitis (AIH) is a rare condition of liver inflammation associated with overactive CD4 cells early in the disease and increasing importance of CD8 cells as the disease progresses. Previous cases of AIH in HIV patients occurred only in those with preserved CD4 counts or with reconstitution of immunity after starting antiretroviral therapy (ART). Here, we present an unusual case of a biopsy confirmed AIH in a patient naive to ART with newly diagnosed AIDS.

Case Presentation: A 42-year-old male with history of Type 2 DM, hypertension, and HIV presented with progressive pruritus and jaundice over several months. He was diagnosed many years ago with HIV with a preserved CD4 count, but unfortunately was lost to follow up. The patient appeared uncomfortable, thin, and jaundiced. Exam revealed scleral icterus, diffuse excoriations, and a distended abdomen with hepatomegaly. Laboratory studies showed CD4 of 94, HIV VL of 349,000, AST 100, ALT 210, T-bili 11.2, D-bili 9.1, INR 1.0, alkaline phosphatase 345, ANA 1:640, anti-smooth muscle positive, and CMV PCR of 203 (normal<200). Liver biopsy showed moderate chronic hepatitis with plasma cells and peri-portal fibrosis, confirming the diagnosis of AIH. CMV tissue stain was negative. After considering the severity of his clinical symptoms, evidence of ongoing liver damage, and risk of fulminant hepatic failure, the patient was started on concurrent Prednisone and ART therapy. He had rapid biochemical and clinical improvement and was discharged by day 3 of treatment with close follow up.

Discussion: Autoimmune diseases are uncommon in AIDS patients. It appears a paradox to have suppressed immunity co-exist with a disease of overactive immune system. Existing research shows that as preferential destruction of CD4 cells in AIDS leads to a pathological CD4/CD8 ratio, it may co-occur in primary CD8 autoimmune diseases such as psoriasis, reactive arthritis, and uveitis. These can even be considered indicators of AIDS, and standard of care includes symptom management and prompt initiation of ART. In this case, we had an ART-naive patient in who we suspect his initial AIDS manifestation was the onset of severe CD8 mediated AIH. This created an interesting diagnostic and therapeutic dilemma as there are currently no well-reported cases or treatment guidelines. After thorough workup was done to rule out other possible etiologies at risk in this population, the decision was made to start therapy based on the low risk for adverse effects and no evidence of corticosteroids being contraindicated in AIDS patients. This case could set future precedent for management of AIH and other more aggressive autoimmune diseases in the immunocompromised patient.
Partial splenic embolization, a Low morbidity alternative option in management of severe ITP

Authors: Banreet Singh Dhindsa, M.D., Yen Cao, M.D., Brian Holloway, M.D., Majed Abuhajir, M.D.

Introduction: ITP (immune thrombocytopenic purpura) is an autoimmune disorder characterized by thrombocytopenia as a primary event or secondary to other causes. In adults, it is more often a chronic disease as compared to children with prevalence of 12 per 100,000 in US.

Severe chronic ITP is defined as platelet count (PC) less than 30,000/ml of blood and results in increased risk of bleeding including intracranial hemorrhage. Current treatment options include immune-suppressive therapy, splenectomy, gamma-globulin infusion and thrombopoietin receptor agonists. The morbidity from chronic immunosuppression and splenectomy has prompted search for an alternative way to manage ITP.

In our case, we explored an alternative modality of treatment known as partial splenic embolization (PSE).

Case Presentation: Our patient is a 49-year-old man who presented with severe isolated thrombocytopenia of 2,000, without other hematological abnormalities. Extensive evaluation was negative for TTP or DIC and for potential secondary causes of ITP such as HIV, HCV underlying autoimmune disorders and lymphoproliferative disorders. Patient responded well to IVIG infusion, however his thrombocytopenia continued to recur few weeks after infusion. The patient subsequently was treated with rituximab with adequate response for one year period. His PC dropped again and he was not responsive to subsequent rituximab and dexamethasone and started having minor bleeding manifestations. Patient still showed adequate response to IVIG infusion indicating that he was a good candidate for splenectomy. We referred the patient to interventional radiology for PSE and 60% of the spleen was embolized. His PC responded promptly within 1 day and remained at 100,000 or higher after 1 year of PSE with no bleeding manifestations. Patient had no Howell-Jolly bodies on his peripheral blood smear indicating no evidence of hyposplenism.

Discussion: Splenectomy became the standard of care in patients with ITP not responsive to steroids over the last several decades. Although majority of patients respond to splenectomy, some patients do not. Intervention is recommended once PC drops below 30,000 as it increases risk of bleeding manifestations including ICH. Primary ITP patients, without identifiable secondary causes are often treated with steroids with subsequent splenectomy if unresponsive. Newer options include IVIG, anti-D Immunoglobulin, thrombopoietin receptor agonists and rituximab infusions. The downside of these therapies are immune suppression and chronic administration of expensive medications.

Splenectomy is a major surgery which results in decreased immunity and increased risk of infections. The alternative option of PSE offers an opportunity to improve the thrombocytopenia while retaining sufficient residual splenic function which is lost in traditional splenectomy.

PSE for ITP management has been reported in limited case series or case reports. More studies need to be undertaken to further evaluate the optimal role of PSE in management of ITP.

References

Neuro-Myelitis Optica: Is There a Role for Prolonged Immunoglobulin Therapy?

Authors: Sailaja Pisipati, Swapna Patel, Paul Park, Biswas Upadhyay, Gurpreet Chahal

Introduction: Neuromyelitis optica (NMO), also known as Devic’s disease is a rare autoimmune, demyelinating disorder characterized by optic neuritis and acute myelitis potentially leading to blindness and paralysis. There is step-wise deterioration due to accumulating visual, motor, sensory, bladder deficits from relapses. Prognosis is grim and associated with high mortality due to neurogenic respiratory failure. We present a case of sero-negative NMO that responded to prolonged immunoglobulin therapy.

Case Presentation: A 43-year-old otherwise healthy lady presented with 2-days of generalized weakness, fatigue and 1-day of acute ascending lower limb paresthesias and motor weakness associated with mild back pain and voiding difficulty. Examination revealed complete flaccid paralysis of lower extremities and truncal musculature with loss of deep tendon reflexes and sensory level at T6. CT Head was normal. MRI spine revealed multi-segmental myelopathic hyperintensities in cervical and thoracic cord consistent with longitudinally extensive transverse myelitis (LETM), which is characteristic of NMO. CSF revealed a cell-count of 937 with lymphocytic-mononuclear pattern. CSF AQP4-Abs, HSV, West-Nile IgG, EBV, Enterovirus, oligoclonal bands were negative. Serology for HIV, HTLV-I/II, Hepatitis panel, EBV, West-Nile Virus, Enterovirus, Coccidioides, Coccidioidomycosis, VDRL was negative. Although ANA was detected (1:160), autoimmune workup for SLE, Sjogren’s, polymyositis, mixed connective tissue disorders, anti-phospholipid syndrome, autoimmune thyroiditis was unremarkable.

Treatment with high-dose methylprednisolone for 5 days followed by 10 sessions of plasmaphoresis resulted in minimal improvement. 5-day course of intravenous immunoglobulin (IVIg) provided additional marginal improvement. Following transfer to acute rehab, she had flare of symptoms and was given a second course of high-dose methylprednisolone and IVIg that resulted in some improvement. Prolonged therapy with IVIg every 4-weeks over the next 12-months coupled with intense physical and occupational therapies resulted in complete recovery of function in her right leg, near-complete recovery in left leg, abdominal and gluteal musculature.

Discussion: NMO should be high on the list of differentials in any patient presenting with clinical and radiological evidence of transverse myelitis. Negative AQP4-Ab cannot exclude the diagnosis. AQP4-Ab–negative patients with LETM and NMO could have alternative diagnoses; it is hence vital that patients with LETM are screened for other causes before being labeled as NMO. Treatment is similar between seropositive and seronegative NMO. First line treatment is high-dose methylprednisolone. For patients unresponsive to glucocorticoids, therapeutic plasma-exchange is recommended. Long-term immunosuppression for preventing recurrent attacks is associated with adverse-effects. Novel biological agents, targeting humoral and cellular markers, are under development to improve long-term management and outcome, and to decrease side-effects. Role of IVIg in treating acute attacks is not well established. Our patient had a remarkable recovery following a second course of IVIg and subsequent monthly injections. Our case highlights that repeated and prolonged treatment regimen with IVIg is well-tolerated and could result in remarkable functional recovery.
Herbal trouble! A Rare Case of an Ayurvedic Diabetes Remedy Causing Lead Poisoning

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Introduction: Complementary and alternative medicine use is as widespread in the United States today as it is across the globe. Its use is assumed to be safe and free from side effects. In addition to organic compounds, Ayurveda uses lead, arsenic and mercury as active elements in its formulations. Cases of lead toxicity have been reported with the use of ethnic remedies when used for the treatment of hemorrhoids, psoriasis and tooth pain. We report a rare case of chronic lead poisoning from an herbal remedy used for diabetes management.

Case Presentation: A 60-year-old Indian male presented with complaints of multiple bouts of squeezing, non-radiating abdominal pain all over his belly for the past 5 years which had worsened in intensity and frequency over the last week. Peaking to an intensity of 10/10 in minutes the pain was often accompanied by nausea but not vomiting, fever, sweats, chills, rashes, flatulence, photosensitivity or urinary complaints. He recognized no aggravating factors or relation to food intake. Ibuprofen and acetaminophen had provided only marginal relief. Extensive testing over the past 5 years including computed tomography scans, ultrasounds and endoscopies had yielded no answers. His other complaints were significant for diffuse vague aches and pains, headaches, constipation, tingling in his feet, recent erectile dysfunction, insomnia and irritability. He was on treatment for diabetes, hypertension and anemia. His medications included: metformin, insulin (glargine & aspart), aspirin, simvastatin, losartan, iron and “Madhumehatakvati” – an herbal supplement for diabetes management from India which he had been consuming for the last 10 years. He lived in a house built in 1985, worked as a researcher at Colgate, did not drink or manufacture his own alcohol or abuse illicit drugs.

His vitals were stable, conjunctivae were pale, and the junction of the teeth and the gums showed discoloration (Burton’s Lines). The abdomen was non-tender with no organomegaly. Both feet showed decreased sensation to monofilament testing.
Lab analysis showed microcytic anemia (Hb 8.8 gm/dl; MCV 78.8 fl.), basophilic stippling on peripheral smear and markedly elevated serum lead (142 mcg/dl) and zinc protoporphyrin levels (718 mcg/dl). Plain abdominal radiograph showed 5 pill-sized radiopaque foreign bodies. He was started on chelation with succimer and poison control was noticed.

Discussion: In Ayurvedic medicine, lead is considered an aphrodisiac and its presence in diabetes preparations may be to counter the impotence associated with diabetic neuropathy. Analyses of some of these remedies have often found lead concentrations thousands of times the FDA’s upper limit of normal for water or candy. With the Ayurvedic market projected to grow at 16% over the next decade, this case questions the need to potentially revisit dietary supplements regulation in the United States.
Pulmonary Hypertension, The Undocumented Complication of Congenital Bronchial Atresia: A Case Report

Authors: Prince Alebna MD, Raghav Chaudhary MD, Hwan Kim MD, Killol Patel MD, Englewood Hospital and Medical Center, Englewood, NJ

Introduction: Congenital bronchial atresia(CBA), marked by complete occlusion of the bronchial tree lumen, is often an asymptomatic benign condition with people who have CBA living well into adulthood unaware they have the condition. Recurrent lung infection is the main complication reported in literature on CBA. This case, however, highlights a potential long-term complication of CBA that was hitherto undocumented.

Case Presentation: A 66 year old female with a history of hypertension, coronary artery disease, atrial fibrillation status post ablation presented with dyspnea on exertion for 2 days. She denied orthopnea, paroxysmal nocturnal dyspnea or ankle swelling. Physical exam was only positive for diminished breath sounds in her left upper lung zone and bibasilar crackles. CT chest with contrast ruled out pulmonary embolism but revealed evidence of right heart strain, interstitial edema and hyperlucent left upper lobe consistent with CBA. Echocardiography showed ejection fraction of 50-55%. Left and right heart catheterization revealed patent coronaries, severe pulmonary hypertension (PH) with a mean pulmonary arterial pressure(mPAP) of 50.5mmHg and pulmonary capillary wedge pressure(PCWP) of 26mmHG. Cardiology, pulmonology and cardiothoracic surgery were involved and a decision was made against surgery. She responded to diuresis and was discharged on the 4th day.

Discussion: CBA is rare and often affects segmental or lobar bronchi. Only a handful of cases of CBA are reported in English literature with most writeups originating from Asia and Europe. With a male preponderance, the incidence is estimated at 1.2 per 100,000 males. CT chest is the most sensitive diagnostic modality. Bronchoscopy is only indicated in doubtful cases. The main finding is hyperlucency which is due to hyperinflation of affected alveoli and compression of adjacent tissues. Hyperinflation results from unidirectional air flow via collateral canals called pores of Kohn and canals of Lambert. This causes ventilation/perfusion mismatch which ultimately causes PH after many years. In the index case, the elevated mPAP was way out of proportion of the high PCWP implying a lung pathology (Type 3 PH) was the likely etiology. Although she had left heart failure, her PH was caused by the chronic VQ mismatch, as no other lung pathology was noted.

Existing literature mostly cites recurrent infections and pneumothorax as the main complications. There is no guideline on the management of CBA and surgery is often reserved for serious complications. This case highlights how CBA, a rare congenital condition causes PH, complicates the diagnosis and management of heart failure. Opinions are divided on surgery in asymptomatic patients. Given the chances of PH later in life, early surgery in children and young adults may be beneficial. More research is needed to guide the management of CBA.
NEW JERSEY CLINICAL VIGNETTE POSTER FINALIST - YUVRAJ CHOWDHURY, MBBS

The Heart Slows On Yellow: Hyperbilirubinemia Induced Bradycardia

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Introduction: In 1877, Murchison commented on jaundice noting, “in a large proportion of cases, the pulse is unusually slow.” Up until 1972, this phenomenon merited mention in medical textbooks. However recent references are limited and it has become an association commonly overlooked.

Case Presentation: A 64-year-old man presented to the emergency department with syncope. Earlier that day, he experienced cramping upper abdominal pain that lasted an hour. He visited an urgent-care clinic where he felt lightheaded before losing consciousness. He had no jerking or oral frothing and recovered spontaneously within a minute.

He was thus sent to the emergency department. On examination, he was lying comfortably in bed. Pulse was 39 beats/minute, regular and normovolemic. Blood pressure was 116/52 mm Hg, evaluation for orthostasis was negative and temperature was 98.30F. His conjunctivae were icteric. S1 and S2 were normal with no murmurs or gallops. Abdominal examination revealed no tenderness.

Serum chemistry revealed aspartate aminotransferase 342 U/L, alanine aminotransferase 470 U/L, alkaline phosphatase 145 U/L and total bilirubin 4.8 mg/dl (direct bilirubin: 3.1mg/dl), a pattern consistent with obstructive jaundice. Electrocardiogram showed new sinus bradycardia (43 beats/min), 1st degree AV block (PR interval 216 ms) and RSR’ in lead V1.

Ultrasonography showed cholelithiasis and a 4mm wide common bile duct without choledocholithiasis confirmed on magnetic resonance cholangiopancreatography. Liver chemistries returned to normal during hospitalization suggesting obstructing choledocholithiasis which had since passed. As the hyperbilirubinemia resolved, the heart rate began to rise. Repeat electrocardiogram 22-hours after the initial showed heart rate of 52 beats/minute, reversal of the 1st degree A-V block (PR interval 190 ms) and persistence of RSR’ in V1. He underwent a laparoscopic cholecystectomy the same admission. At discharge, his resting heart rate was 82 beats/minute and total bilirubin was 0.9 mg/dl. His electrocardiogram at outpatient visit 2 months later showed a heart rate of 71 beats/minute, PR interval of 134 ms and unchanged RSR’ in Lead V1.

Discussion: The etiology of syncope was likely sinus and A-V nodal dysfunction secondary to hyperbilirubinemia. Joubert et al ligated bile ducts of rats and found that post- ligation, the mean heart rate decreased from 374 (SD= 21) beats/min to 348 (SD=11) beats/min (P<0.0005). Electrocardiogram of these rats showed a spectrum from bradycardia to varying degrees of A-V block. Bogin demonstrated similar mechanisms by adding the serum of rats whose bile ducts had been ligated to the cultured myocardium. The deoxycholate and cholate rich serum decreased heart rate and led to early cessation of contractions. A literature review reveals this association only holds true with obstructive jaundice. This case highlights the cardio-toxic properties of bilirubin and its potential to induce nodal dysfunction. Indeed, like a model citizen, the heart too slows on yellow!

References

NEW JERSEY CLINICAL VIGNETTE POSTER FINALIST - AHMAD DAMATI

A very rare case of severe thyrotoxicosis presenting as refractory torsades de pointes.

Authors: Ahmad Abuarqouba, Ahmad Damatia, Firas Qaqa a, Fayez Shamoona, Department of Cardiology, Saint Joseph’s Regional Medical Center, Paterson, NJ

Introduction: Acquired torsades de pointes seen commonly in association with electrolyte disturbances and medication. Hypothyroidism is well known to be associated with torsades but it is very rarely seen with thyrotoxicosis.

We are describing the first case in literature of Torsades associated with thyrotoxicosis treated by achieving euthyroid status.

Case Presentation: 53 years old female with unknown past medical history was admitted through ER with 1-week history of increased shortness of breath associated with palpitations, orthopnea, PND and bilateral lower limb edema. History of nausea, occasional vomiting, diarrhea and non-intentional weight loss. Physical examination was remarkable for an irregular-irregular pulse of 170, palpable thyroid, bibasilar crackles and bilateral lower limb pitting edema.

Upon further work up the patient was found to have an elevated FT4 and T3, very low TSH, elevated BNP and mild hypokalemia, EKG showed Atrial fibrillation with RVR, CXR showed pulmonary vascular congestion, neck ultrasound showed multiple thyroid nodules, echocardiogram showed an EF of 25% with global hypokinesis.

The patient was started on methimazole, propranolol and diuresis, with slight improvement in the clinical status, since the patient was still in Afib, TEE and cardioversion was done and the patient reverted to sinus rhythm; Post cardioversion EKG showed sinus rhythm with prolonged QT interval.

1 day post cardioversion the patient had recurrent episodes of polymorphic ventricular tachycardia followed by ventricular fibrillation (telemetry review showed R on a T phenomenon, was defibrillated 6 times, at that time all electrolytes was within normal limits, and no offending medications were found, so the decision was made for transvenous pacemaker insertion and over-ride pacing which prevented further episodes, at that time right and left heart catheterization was done which showed normal coronaries in addition to normal hemodynamic data. Upon a wean off the trial of the backup pacing the patient had further episodes of polymorphic ventricular tachycardia, the patient was kept on back up pacing, and upon achieving euthyroid status within few weeks no further episodes were noted, and the patient was asymptomatic upon outpatient follow up.

Discussion: Out of 300 000 sudden cardiac death in the US, around 5% was attributed to Torsades.

Upon literature review Several case reports of monomorphic ventricular tachycardia was associated with thyrotoxicosis due to underlying electrolyte disturbances, two case seen in literature in which thyrotoxicosis progressed into polymorphic ventricular tachycardia one of them was associated with stress cardiomyopathy.

Torsades de pointes is a very rare manifestation of thyrotoxicosis as hyperthyroidism rarely prolonged the QT interval, this is the first case in literature that links hyperthyroidism with torsades in the absence of any other underlying cause, once achieving the euthyroid status torsades was suppressed.
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NEW JERSEY CLINICAL VIGNETTE POSTER FINALIST - ESEOSA EDO-OSAGIE, MD

SPLENIC SEQUESTRATION CRISIS AS AN INDEX MANIFESTATION OF HETEROZYGOUS HEMOGLOBINOPATHY IN AN ADULT

Authors: Eseosa Edo-Osagie MD, Hisham Hakeem MD, Manoj Rai MD, Emmanuel A. Adomako MD, Maxwell Janosky MD.

Introduction: Acute splenic sequestration crisis (ASSC) is a well-recognized complication in children with sickle cell disease (SCD) but is a rare manifestation in adults with sickle-beta thalassemia (S-β thalassemia). We report a case of ASSC as an index manifestation of HbS β-thalassemia.

Case Presentation: A 20-year-old Guyanese female of Indian descent presented with a five-day history of fever and left upper quadrant abdominal pain, associated with abdominal distension. Physical examination was remarkable for fever of 103°F, conjunctival icterus, left upper quadrant tenderness, and splenomegaly. Laboratory data revealed hemoglobin (Hb) of 6.5g/dl, mean corpuscular volume (MCV) 64.8fl, mean corpuscular hemoglobin (MCH) 22.2pg and an elevated leukocyte count of 14.72 x 10^9 with a left shift. Lactate dehydrogenase (LDH) was elevated (3420 u/l) with decreased haptoglobin <15. Reticulocyte count was 7.5%. Liver function test revealed hyperbilirubinemia. Peripheral blood smear showed markedly hypochromic microcytic red blood cells with target cells and increased reticulocytes. Computed tomography (CT) scan of the abdomen with contrast showed massive splenomegaly (22 cm), consisting of circumferential peripheral and centrally diffuse infiltrative cystic attenuation within the parenchyma. Data was consistent with an acute hemolytic anemia. Coomb’s test, osmotic fragility test and G6PD assay were negative. Sickling test done was positive, Hb electrophoresis revealed sickle cell trait (AS) with Hb percent consisting of HbA-48%, HbS-26.8% and Hb F-24.0% with normal HbA2 (1.2%). DNA test for beta globin gene mutation was pending. She was transfused 2 units of packed red cells, and received empiric antibiotics and supportive care, however, symptoms persisted therefore splenectomy was performed. Pathologic examination of the spleen demonstrated massive splenomegaly with peripheral serpiginous yellow infarcts, surrounded by acute inflammation and small capillaries plugged with sickle cell shaped red blood cells consistent with splenic sequestration. Postoperatively, there was defervescence of fever and improvement of Hb to 10.6g/dl, and she was discharged home. At follow-up two weeks later, she was completely asymptomatic. DNA test revealed beta-globin mutations consistent with sickle-beta-thalassemia.

Discussion: Acute splenic sequestration syndrome and acute splenic infarction are sequelae of sickle hemoglobin disorders. According to various case reports the association of S-β thalassemia with splenic sequestration crisis is uncommon [1]. There are no apparent precipitating factors for S-β thalassemia associated ASSC in adults [2], even though some hypothesize that high altitude and infections can precipitate the crisis. Transfusion of blood products and supportive care can reduce the severity of the crisis. Splenectomy can be considered in cases with recurrent splenic sequestration crisis, or those with double heterozygous sickle hemoglobinopathies with acute splenic sequestration syndrome who fail to show clinical improvement [3]. This case highlights the wide variety of clinical phenotype encountered with S-β+ thalassemia. A high index of suspicion should be maintained to minimize unnecessary testing and ensure prompt and appropriate management.

References

Elizabethkingia meningoseptica bacteremia in a patient with mucinous biliary adenocarcinoma

Authors: Richard A. Greendyk MD, Ian J. Mahoney MD, Emilia A. Hermann MD MPH

Introduction: *Elizabethkingia meningoseptica* (*E. meningoseptica*, previously classified as *Chryseobacterium meningosepticum* and *Flavobacterium meningosepticum*) is a Gram-negative bacillus, historically associated with meningitis in premature neonates. It is increasingly recognized as a cause of multi-drug resistant healthcare-associated infections in both immunocompetent and immunocompromised adults.

Case Presentation: A 50 year old obese woman with a history of uterine fibroids status post hysterectomy was admitted to the General Medicine service for several weeks of right-sided abdominal pain and jaundice. Labs showed direct hyperbilirubinemia (total bilirubin 8.0, direct bilirubin 6.7, AST 344, ALT 607, alkaline phosphatase 425). CT scan of the abdomen/pelvis revealed a 6.7x3.6cm mass centered around the gallbladder with intrahepatic extension, biliary ductal dilatation, and periportal lymphadenopathy. Fine-needle aspiration of the mass demonstrated mucinous adenocarcinoma with signet-ring cell features, thought to be of biliary origin. Endoscopic retrograde cholangiopancreatography (ERCP) and partial hepatectomy were attempted to decompress the biliary tract, but could not be performed due to tumor burden. Interventional radiology performed left and right percutaneous biliary drain placement, with initial improvement in hyperbilirubinemia from total bilirubin 17.6 (direct bilirubin 15.9) to nadir total bilirubin 5.9 (direct bilirubin 4.7). Despite two biliary drain revisions, the patient developed worsening hyperbilirubinemia in the following three weeks. Repeat CT abdomen/pelvis demonstrated progression of disease with diffuse biliary ductal dilatation not amenable to percutaneous drainage. The patient subsequently developed fever to 38.6C, leukocytosis (WBC 24.6 with 94.0% neutrophils), and total bilirubin elevation to 12.0. Blood cultures grew multi-drug resistant *E. meningoseptica* identified in two of two culture sets less than twenty-four hours after cultures were obtained. Urine and respiratory cultures were negative and chest x-ray was clear. The patient was started on empiric meropenem and minocycline. Blood cultures remained persistently positive with *E. meningoseptica* despite antimicrobial therapy. The patient developed septic shock, and family elected to pursue a palliative course. The patient died of multi-organ failure secondary to septic shock from *E. meningoseptica* bacteremia.

Discussion: While *E. meningoseptica* bacteremia has been identified in patients admitted from the community, it is typically a healthcare-associated infection. Although it is unclear how *E. meningoseptica* is contracted in isolated cases, contaminated saline, chlorhexidine gluconate, and lipid solutions have been identified as sources of infection in outbreaks. Several case reports also document the development of *E. meningoseptica* cholangitis after ERCP for cholelithiasis/choledocholithiasis. Cholangitis with *E. meningoseptica* is generally associated with good outcomes if biliary obstruction can be relieved and appropriate antibiotics are given, although bacteremia can be fatal. This case highlights the severity of *E. meningoseptica* cholangitis in patients with unrelied biliary obstruction as in this patient with inoperable malignancy. Further investigation is needed to identify mode of transmission and optimal infection control procedures in isolated cases that develop after biliary tract instrumentation.

References

ATYPICAL HEMOLYTIC UREMIC SYNDROME COMPlicated WITH ESRD

Authors: Holguin S

Introduction: Hemolytic uremic syndrome is a microvascular thrombosis associated with thrombocytopenia, hemolytic anemia and acute kidney injury. The disease is further classified as typical (85-90%) or atypical (15-10%) depending on the etiology. The former is related with Shiga toxin-producing Escherichia coli (STEC); meanwhile the latter could be precipitated by other types of infections (predominantly S. pneumoniae, influenza A, HIV), drug toxicity, autoimmune diseases or genetic disorders. The annual incidence of atypical hemolytic uremic syndrome (aHUS) in adults is approximately 2/1,000,000 versus 3.3/1,000,000 in patients under age of 18. The complement system, especially the alternative pathway, is the cornerstone of the pathogenesis of aHUS. Either genetic mutations in complement factor H and the CD46 or uncontrolled C3 convertase activity, precipitated by any of the above-mentioned factors, can lead to deposition of complement proteins in the vessel and subsequent prothrombotic state. Unlike patients with typical HUS in which 70% of the patients completely recover their renal function, up to 50% progress to end-stage renal disease (ESRD).

Case Presentation: This case describes the hospital course of a 76-year-old woman who presented with dyspnea and oliguria. Creatinine was 3.6 mg/dL upon admission, and she eventually developed anemia and thrombocytopenia. ADAMTS-13 was moderately decreased, and the antibody panel and complement levels were unremarkable. Due to volume overload, the patient was started on renal replacement therapy and a renal biopsy was performed demonstrating thrombotic microangiopathy with diffuse positive staining of C5b-9 within glomeruli and small arterial vessels. Her hospital course was complicated with a pericardial effusion requiring pericardial window. After preliminary biopsy results were obtained, treatment with eculizumab (monoclonal antibody against complement factor 5) began. Patient still requires three times per week hemodialysis and additional treatment with C5 inhibitor.

Discussion: Atypical hemolytic uremic syndrome is a very rare disease which presents with the triad of thrombocytopenia, nonimmune hemolytic anemia and acute kidney injury. This type of microvascular thrombosis can progress to ESRD in 50% of the cases. Its etiology varies from infectious causes to genetic disorders, all of them having in common the dysregulation of the alternative pathway of complement. Several studies have demonstrated gradual recovery of the kidney injury with long-term C5 inhibitor such as eculizumab.

References

Spontaneous Regression of an High Grade Diffuse Large B-Cell Lymphoma of the Liver

Authors: Yi Jiang, Resident Physician, Department of Internal Medicine, Rutgers New Jersey Medical School, Newark, NJ., Victor Chang, Attending Physician, Section of Hematology/Oncology, VA New Jersey Health Care System, East Orange, NJ.

Introduction: Spontaneous regression (SR) of cancer is a condition characterized by partial or complete disappearance of a malignant tumor in the absence of treatment or in the presence of therapy considered inadequate. Although not uncommon in low-grade lymphoma, SR of diffuse large B cell lymphoma (DLBCL) is extremely rare. Here we report a unique case of untreated DLBCL of liver with spontaneous remission.

Case Presentation: A 64-year-old man with history of chronic hepatitis C virus (HCV) infection presented to the Emergency Department for a 2 week history of right upper quadrant abdominal pain. Workup led to triple phase abdominal computed tomography (CT) that showed a 6 cm right hepatic mass and borderline lymphadenopathy. Liver core biopsy pathology showed diffuse large B cell lymphoma CD20 and BCL6 positive, with Ki-67 greater than 90%. Further staging work up with bone marrow exam and positron emission tomography (PET) CT confirmed stage IVA DLBCL, not otherwise specified (NOS) with high-grade histologic features limited to the liver. Based on the International Prognosis Index system, patient was considered to be in the high-intermediate risk group. The patient’s treatment was delayed and a PET CT scan repeated at 3 months from initial diagnosis in preparation for treatment showed partial regression of the liver lesion with eventual complete resolution of the hypermetabolic status. The patient has been closely followed and the most recent PET CT at 34 months follow up was normal. The patient continues to do well.

Discussion: In previous case reports, the majority of cases of SR were for DLBCL located in head/neck, breast and stomach. Patients with primary high grade lymphoma of the liver have a poor prognosis. This case demonstrated a SR of primary liver DLBCL with aggressive clinical and histopathologic features. This observation suggests immune mechanisms can be important in affecting the outcomes of high grade lymphoma. Uncovering the underlying mechanism of SR may shed light on new therapeutic strategies to improve outcomes for future patients.

References

Pylephlebitis Presenting as Fever of Unknown Origin

Authors: Philip Kanemo, Mariam Saand, Sabena Ramsetty

Introduction: Pylephlebitis, infective suppurative thrombosis of the portal vein, is typically associated with intraabdominal infections. It is often characterized by fever, abdominal pain, and polymicrobial bacteremia. We present a case of pylephlebitis presenting as indolent fever of unknown origin (FUO).

Case Presentation: An 82-year-old man with diabetes presented with a four-month history of fevers, drenching night sweats, thirty-pound weight loss, and vague right-sided abdominal pain. Outpatient CBC and chemistries were unremarkable, blood cultures were negative, and CT scan of chest and abdomen without contrast revealed possible lung infiltrates. Patient received one week of Levofloxacin for possible pneumonia with no clinical improvement. His last colonoscopy at age 52 was normal. He denied travel, sick contacts or tick exposures. He was admitted for further workup.

On admission, he appeared weak and diaphoretic. He was febrile to 100.5. Physical exam was only remarkable for mild right upper quadrant tenderness. Labs revealed leukocytosis of 14.74 k/ul, hemoglobin 10.6 g/dl and platelets 250 k/ul. Electrolytes and liver function tests were normal. Gallium scan was ordered and was non-diagnostic. CT abdomen/pelvis was repeated with contrast revealing extensive portal vein thrombosis. Hypercoagulable workup was negative. Fusobacterium was isolated from one of five sets of blood cultures drawn during hospitalization. Metronidazole was started and patient defervesced within 48 hours. He was discharged home with 6 weeks of oral Metronidazole as well as anticoagulation.

Discussion: Pylephlebitis is a rare but serious condition which may complicate intra-abdominal conditions including diverticulitis, appendicitis, peritonitis, and rarely Crohn’s disease. It can be life-threatening if it goes undiagnosed. Fusobacterium is a slow-growing anaerobe, and there are several reported cases documenting its association with thrombophlebitis of the portal vein as well as internal jugular vein thrombophlebitis (Lemierre’s syndrome). The organism itself is thought to have thrombogenic properties. Management of pylephlebitis typically involves a prolonged course of pathogen-targeted antimicrobials in addition to several months of anticoagulation.

Pylephlebitis should be considered in cases of FUO presenting with vague abdominal pain. Our case illustrates the importance of contrast imaging of the abdomen/pelvis when evaluating a patient for FUO. This should be done before nuclear imaging or invasive diagnostic tests. We also emphasize the importance of obtaining multiple blood cultures in efforts to isolate fastidious organisms such as Fusobacterium.

References

Cardiomyopathy - A diagnostic dilemma

Authors: Harsh Mehta, Saad Amin, Rohma Shaikh, Sargis Khoobiar

Introduction: Peripartum cardiomyopathy (PPCM), and Tako-tsubo cardiomyopathy (TTCM) are uncommon forms of cardiomyopathy, with unknown etiologies demonstrating similar phenotypic features of reduction in left ventricular systolic function. We report a case of cardiomyopathy in a young female with overlapping features of PPCM and TTCM.

Case Presentation: A 36-year-old Caucasian female, with a history of pre-eclampsia and twin birth during first pregnancy was admitted for caesarean section. Post-operative course was complicated by post-partum hemorrhage, requiring exploratory laparotomy and evacuation of 1.5 liters hematoma. She remained hypotensive and tachycardic intra-operatively and post-operatively, requiring blood transfusions, fluid and pressor support. She remained intubated and mechanically ventilated post-operatively. EKG was significant for new ST segment depressions in anterior leads with T wave inversions in inferior leads accompanied by elevated cardiac enzymes (troponin-T 0.556, CKMB-16.3). Chest-XR was suggestive of mild congestion. Echocardiogram (ECHO) showed left ventricular ejection fraction (LVEF) of 30-35%, left ventricular end-diastolic dimension (LVEDd) 2.9cm/m2 with severe global hypokinesis except for a hyperkinetic apical segment. Patient was started on carvedilol, lisinopril and atorvastatin, anti-coagulation was avoided due to bleeding. Cardiac catheterization showed normal coronaries without evidence of stenosis or dissection. Over the next 4 days, her vital signs and hematological parameters stabilized, and she was discharged with continuation of the above-mentioned medications. ECHO at six-week follow-up was suggestive of resolution of the wall-motion abnormalities with LVEF 55-60%.

Discussion: PPCM is defined as the development of dilated cardiomyopathy from the last month of pregnancy through the first 5 months after delivery, when other causes of heart failure have been excluded. Typical ECHO findings include globally decreased contractility with LVEDd≥2.7 cm/m, LVEF ≤45% and M-mode fractional shortening ≤30%. Our patient met the diagnostic criteria and had multiple associated risk factors including older maternal age, multiparity, multifetal pregnancy and pre-eclampsia. TTCM is a cardiomyopathy characterized by a transient reduction in LVEF, associated with emotional or physical stress. Although initially reported as apical ballooning syndrome, other forms have subsequently been recognized including a reverse variant characterized by basal hypokinesis with or without apical hyperkinesis. Apical ballooning occurs more commonly in postmenopausal women, whereas the reverse variant is seen in younger females, and is invariably associated with a stressor. Our patient showed features of reverse variant of TTCM probably precipitated by sudden loss of intravascular volume secondary to delivery and intra-abdominal bleeding. This case is unique in that our patient demonstrated features of both PPCM and reverse variant of TTCM simultaneously. While prolactin, 16kDa prolactin and cathepsin D mediated oxidative stress is theorized to be involved in PPCM, TTCM is thought to be caused by catecholamine surge and coronary vasospasm. In conclusion, we propose that further research is required to definitively distinguish these two etiologies of dilated cardiomyopathy.

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Diagnosis and Management of Aortitis Secondary to Takayasu’s Arteritis

Authors: Natalie Millet D.O., Magdalena Szulc M.D., Mitul Kanzaria M.D.

Introduction: Aortitis, the inflammation of the aorta, is an extremely rare disease with an incidence of 1-2 people per million in the United States. Aortitis has been demonstrated to involve both the thoracic and abdominal aorta. In some instances, the aorta is exclusively affected however, in cases secondary to vasculitis, the inflammation may extend to the major branches of the aorta. We discuss the diagnosis and management of a case of abdominal aortitis secondary to Takayasu’s arteritis.

Case Presentation: A 45 year old male presented to the emergency department with the complaint of severe bilateral testicular pain. His symptoms began as low back pain following what he describes as a viral illness that occurred approximately three weeks prior to presentation. His back pain and abdominal pain progressively worsened and began radiating to his testicles prompting him to seek further care. On presentation to the emergency department, a CT of the abdomen was ordered to evaluate for nephrolithiasis however, a markedly inflamed abdominal aorta with findings suspicious for aortitis was instead discovered. A CT Angiogram of the chest, abdomen and pelvis was subsequently performed which again demonstrated abnormal enhancing tissue surrounding the abdominal aorta spanning a total of 6 cm. The inflammation began distal to the renal arteries and extended to the bifurcation of the iliac arteries. Also demonstrated was an ascending aortic aneurysm measuring 4.5 cm at the level of the pulmonary trunk.

Intravenous steroid therapy was initiated immediately on admission for treatment of aortitis. Significant lab findings included an elevated erythrocyte sedimentation rate (ESR) and C-reactive protein with an ESR of 63 and CRP of 53. Rheumatologic studies including Perinuclear antineutrophil cytoplasmic antibody (P-ANCA), Cytoplasmic antineutrophil cytoplasmic antibodies (C-ANCA), Antinuclear Antibody (ANA), Rheumatoid Factor and IgG4 all returned negative. Blood cultures and syphilis testing returned negative as well. An echocardiogram was performed to rule out endocarditis as the etiology of the patient’s aortitis; an ejection fraction of 40-45% with stage II diastolic dysfunction was discovered. The patient subsequently underwent cardiac catheterization with findings of coronary artery ectasia involving the left main, left anterior descending, right coronary and circumflex arteries.

Discussion: In addition to meeting the American College of Rheumatology 1990 Criteria for diagnosis of Takayasu’s arteritis, vascular abnormalities involving both medium and large vessels were discovered supporting the diagnosis of Takayasu’s arteritis. Management was a challenge given the paucity of literature regarding treatment of aortitis. After six months of steroid therapy and methotrexate, the patient’s vasculitis appears to be in remission as repeat imaging demonstrates resolution of the inflammation surrounding the abdominal aorta. ESR and CRP have also normalized after therapy.

References


NEW JERSEY CLINICAL VIGNETTE POSTER FINALIST - NEVEEN E MOSTAFA, MD

Dangers of PEG placement in morbid obesity

Authors: Neveen Mostafa, Hussein Hussein, Rabeea Nazir, Henok Tumebo, Farnoosh Farshidi, Babak Jamasian

Introduction: Morbid obesity is a problem that has become globally widespread, and may be an aggravating factor to many medical problems. Percutaneous Endoscopic Gastrostomy (PEG) tubes are frequently used in patients with swallowing dysfunction but can be lethal in morbidly obese patients.

Case Presentation: A 49-year-old morbidly obese male with BMI of 40 was in the critical care unit on mechanical ventilation for respiratory failure, and feeding was started via OGT for 48 hrs which provided the required calories. After failure of multiple attempted extubations, patient underwent tracheostomy and percutaneous gastric tube placement, which was placed by open surgical approach. Feeding was switched to PEG 6 hrs after the tube was tested and confirmed functioning well. Feeding started with gradual advancement and reached the goal next day, but after 2 days he developed fever of 101 F with elevated WBC. Sepsis protocol was initiated and patient continued to deteriorate and required pressors with no obvious source of infection. His abdominal examination revealed soft abdomen; no obvious skin changes or erythema; however, the patient was mechanically ventilated and sedated. While assessing his PEG, there was no leak or resistance in the flow inside the tube, however he was still spiking fevers and in septic shock despite negative blood cultures. CT abdomen and pelvis revealed a large amount of gas in the anterior abdominal wall suggestive of necrotizing fasciitis with the gastrostomy tube mislocated in the abdominal wall. Patient was taken immediately to operating room for debridement, and the collected abscess was widely opened and drained. Suddenly, the patient lost the pulse, and CPR was initiated for greater than 20 minutes with multiple rounds of epinephrine, atropine, and sodium bicarbonate but unfortunately was unsuccessful.

Discussion: PEG placement may have increased challenges in morbidly obese patients due to thick subcutaneous tissue and associated limited examination secondary to body habitus. Indications for PEG placement should be carefully considered in every obese patient before placing, and use of longer tubes with frequent and careful physical examination by skilled personnel is recommended. Moreover, abdominal imaging should be considered in obese patients as early as possible especially if there are signs of sepsis.
How Low Can You Go? Severe Hyponatremia With a Sodium of 94 mg/dL Complicated By Retroperitoneal Hemorrhage

Authors: Nikesh Patel, DO, Sophia Kwon, DO, Abraham Lo, DO

Introduction: Hyponatremia is commonly defined as a serum sodium concentration below 135 meq/L. It is the most common electrolyte disorder in medicine that can range from a myriad of different etiologies ranging from drug induced to SIADH. Management of hyponatremia is determined by level of sodium, symptomology, and etiology. Severe hyponatremia is classified as sodium levels of less than 120 with symptoms including confusion, seizures, and coma. Management is difficult as correcting too rapidly can cause severe neurological impairment, most notably causing osmotic demyelination syndrome.

Case Presentation: An 83-year-old male with a past medical history of Diabetes Mellitus Type 2, benign prostatic hyperplasia, and hypertension presented with nausea and vomiting for three days. Oral intake was poor, but he reportedly drank 18 glasses of water the day prior to admission and was recently started on hydrochlorothiazide for hypertension. Physical exam was benign, but serum sodium was 94 mg/dL. Patient was monitored in the intensive care unit for sodium correction with fluid restriction, and concomitant 3% hypertonic saline and Desmopressin. Hypertonic saline was administered as 50 cc/hr boluses with 2 mg of desmopressin q6h for the first 3 hospital days until Na was 105 mg/dL.

Hospital course was complicated by retroperitoneal hemorrhage and hypovolemic shock secondary to enoxaparin use. A resultant acute tubular necrosis further complicated the initial management of fluid restriction. Patient was transfused 3U of packed red blood cells, and bleeding managed conservatively. Patient was discharged without long-term neurologic or renal complications and a sodium of 136 mg/dL.

Discussion: Literature review presents only one other case of a more severe hyponatremia with a sodium of 87 mg/dL. In contrast, our patient had no long term neurological complications nor renal replacement requirements. To our knowledge, we present the first case of severe hyponatremia complicated by the need for multiple blood transfusions and fluid boluses with a successful outcome of treatment with the infrequently used strategy of concomitant hypertonic saline and desmopressin. The extreme hyponatremia in this patient was multifactorial secondary to severe ongoing sodium and water loss from vomiting, large amount of water intake, recent initiation of thiazide diuretic, low dietary solute intake, urinary retention, and SIADH.

Management was further complicated as patient developed retroperitoneal hemorrhage secondary to DVT prophylaxis and needed blood transfusion. Blood transfusion can often times lead to further electrolyte imbalance affecting such entities such as sodium. This case demonstrates the medical management of sodium level that often times can lead to death and further complicated with bleeding and necessity of blood transfusion, which was effectively treated and had the patient suffer no long term neurologic or renal injury.
Recurrent Confusion- A rare case of non-traumatic pneumocephalus

Authors: Chaitali Patel; Ratan Chaturvedi; Laeeq Butt; James K. Liu

Introduction: Pneumocephalus is defined as pathological collection of gas within the cranial cavity accumulating in the epidural, subdural, subarachnoid, intraventricular, or intraparenchymal compartments. Spontaneous, non-traumatic pneumocephalus is an uncommon condition. The majority of pneumocephalus cases are due to trauma. The typical causes are barotrauma, Valsalva maneuvers, bacteremia and air cell hyperpneumatization. The developmental mechanism is mainly based on two factors: a reduction in intracranial pressure and the presence of a defect in the dura. It is caused by either a ball-valve mechanism that allows air to enter but not to exit or by CSF leakage which creates a negative pressure with subsequent air entry.

Case Presentation: We present a rare case of a 67 year old male brought to the hospital after his family found him incontinent of urine with “odd behavior,” headache, confusion while driving and inability to follow simple directions. The patient recently had severe upper respiratory tract infection with severe coughing and forceful sneezing. The patient only answered “yes” and “no” to questions and seemed extremely confused. No history of any trauma or surgery elicited. Medical history was remarkable for atrial fibrillation on rivaroxaban, alcohol abuse and hypertension. Head CT showed pneumocephalus in the subdural spaces, subarachnoid spaces, and intraventricular compartments. There was also a foci of air in the right temporal horn of the lateral ventricle, adjacent to the right mastoid process. The patient was discharged after a few days of observation and resolution of pneumocephalus, with follow up as an outpatient. Within a few days of discharge, he was brought back to the hospital with similar symptoms and repeat CT scan showed a significant recurrence of pneumocephalus in the subarachnoid and intraventricular distribution. CT temporal bones showed a thin tegmen of the right middle fossa skull base, suggesting a possible source of dehiscence of the right mastoid roof. CT cisternogram was done, which did not show any CSF leak. It was suspected that the pneumocephalus was arising from the thin tegmen and therefore a middle fossa craniotomy was performed. A small encephalocele associated with a skull base defect in the middle fossa floor was identified with removal of encephalocele and repair of the skull base defect. Postoperatively, pneumocephalus resolved with improved mental status and no further recurrence on imaging at 3 months follow-up.

Discussion: The majority of cases of pneumocephalus are secondary to trauma or medical intervention. In our patient the occurrence could be due to the severe coughing associated with forceful sneezing. To our knowledge there are around 10 cases reported in literature of spontaneous pneumocephalus. Whether to opt for surgery or conservative therapy depends on etiology as well as severity. This case highlights the need for surgical treatment in patients with a “recurrent pneumocephalus”, as only observation may cause severe complications.
An Interesting Presentation of Rare Cervical Cancer

Authors: Aditi Saha MD, Shanojan Thiyagalingam MD, Samar Hameed MD, Sunil Sapru MD

Introduction: Neuroendocrine carcinoma of the cervix is a rare but aggressive form of cervical cancer, accounting for only 1-1.5% of all cases, with small cell carcinoma being the most common type. The biology of these cancers is different from the more commonly seen cervical cancers, with early lymphatic spread and distant metastases usually present at diagnosis. There are no established treatment guidelines and the prognosis remains poor. We describe a patient with high grade small cell cervical cancer who presented with a left groin mass and severe hypercalcemia.

Case Presentation: A 68 year-old woman with no significant medical history presented to the emergency department with complaints of fatigue, 30 pound weight loss, and worsening painful left groin mass for the past 8 months. Physical exam was notable for a 10x12cm well-demarcated, firm, tender mass on her left proximal medial thigh. Her labs were significant for calcium of 14 mg/dL. Hip x-ray revealed lytic lesions in the left superior/inferior ramus of pelvis; MRI pelvis showed a 17x14cm osseous mass at the left pubic bone with invasion of surrounding soft tissue as well as another 11x9cm mass at the uterine body. Cervical biopsy demonstrated small cell carcinoma, staining positive for synaptophysin, CD56, cytokeratin AE1/AE3, cam5.2, p40; Ki67 proliferation index of 70%. Chemotherapy with Carboplatin and Etoposide was initiated. Her clinical condition deteriorated, she developed septic shock, and passed away one week later.

Discussion: Neuroendocrine tumors, usually of lung or GI origin, are rarely also seen in the female genital tract. Small cell carcinoma of the cervix, a poorly differentiated neuroendocrine cancer, typically presents in fourth to sixth decade of life. No definitive risk factors have been identified but there is a strong correlation with HPV type 18. Initial presentation includes abnormal vaginal bleeding but can be varied due to its propensity to metastasize early to bone, brain or liver. Paraneoplastic syndromes are not common but rare cases of SIADH and Cushing’s have been reported. Our patient is interesting as she presented with severe hypercalcemia. Histopathology remains the gold standard for diagnosis. Immunohistochemical staining for synaptotophysin, chromogranin, CD56, and neuron specific enolase is used. Presence of any two of these establishes the diagnosis. The 5 year overall survival is 30 to 46%, compared to 65% for adenocarcinoma and squamous cell carcinoma, while in advanced stage it is only 0 to 15%. Management has remained a challenge because of the rarity of this cancer, making it difficult to ascertain a target population to conduct clinical trials. Platinum based chemotherapy and etoposide have been found to be significantly beneficial. Early recognition is crucial and current guidelines for preventative cervical cancer screening are helpful. Further research into management and treatment strategies is needed.

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An Atypical Case of Diabetic Keto-Alkalosis with Severe Hypernatremic Hyperosmolarity in a Patient with Amphetamine Abuse

Authors: Khushpreet Saini, MD, Sridhar Nambi, MD

Introduction: Acid base/metabolic disturbances have a complex interplay in Diabetic ketoacidosis (DKA). Substance abuse in DKA is associated with more severe metabolic acidosis with higher acidosis-ketosis gap.1 Severe hypernatremia and alkalosis is rare in DKA but was exhibited in our case.2,3

Case Presentation: A 31-year-old female with Type1 diabetes and schizophrenia was brought to emergency after she was found to be minimally responsive at home. She had abdominal pain and polyuria since few days and wasn’t compliant with her medications. There was no history of vomiting, diarrhea, alkali ingestion, laxative or diuretic use. On admission, she was severely dehydrated, obtunded and only responsive to painful stimuli. Initial biochemical findings: serum glucose 1323, serum sodium 135, corrected sodium 154, bicarbonate 32, serum osmolality 379, beta hydroxybutyrate 62.56 mg/dl, anion gap 30 and delta gap of 24. Venous blood gas showed: pH-7.40, pCO2-62, pO2-51, HCO3-38.4. She was adequately managed with intravenous fluids and insulin infusion. Patient was fully awake and alert and anion-gap closed within 24 hours of admission. Serum sodium initially peaked to 163 but normalized over the next 36-42 hours. Of note, she was admitted with HHS (Hyperglycemic Hyperosmolar State) 3 months before and 1 month after this admission. Urine drug screen was positive for amphetamines in all 3 admissions.

Discussion: Our patient exhibited mixed metabolic acidosis (elevated anion gap, ketonemia), metabolic alkalosis (elevated bicarbonate, elevated delta gap) and compensatory respiratory acidosis. Osmotic diuresis due to glycosuria and amphetamine abuse (via hyperthermia, excessive sweating, inadequate fluid intake) led to severe volume depletion and bicarbonate reabsorption in our patient leading to severe contraction alkalosis. Hypokalemia also contributes to development and maintenance of alkalosis. Studies have shown that methamphetamine use leads to elevated plasma cortisol and corticosterone levels (greater in females than males) via hyper-activation of the hypothalamic-pituitary-adrenal axis.4 3,4-Methylenedioxymethamphetamine (MDMA)/Ecstasy use has been shown to cause an acute increase in cortisol levels of around 150% in sedentary humans and 800% in MDMA using dance clubbers.5 Excess glucocorticoids promote H+ secretion from kidneys via mineralocorticoid receptors contributing to alkalosis.6 DKA/HHS patients usually present with eunatremia/hyponatremia. Adult schizophrenics who receive psychostimulants have also been shown to develop polydipsia and hyponatremia. Ecstasy intoxication can precipitate both DKA and hyponatremia in Type1 diabetics.7 Cases of combined DKA/HHS and hypernatremic hyperosmolarity have been reported due to huge intake of sugar-rich carbonated carbohydrate beverages to quench thirst especially in pediatric population.3,8 Serum sodium and plasma osmolarity shouldn’t fall more than 1-2 meq/hour and 3 mOsm/Kg/hour respectively in such cases to prevent the development of cerebral edema. There is frequent association between decompensated diabetes and illicit drug use. Substance abuse should be suspected in young Type1 diabetics presenting with recurrent hyperglycemic emergencies especially with atypical biochemical features.9

References


Pulmonary Tumor embolism from Cholangiocarcinoma

Authors: Shrey Shah MD, Ahmed Mohammed MD, Abdur Shad MD

Introduction: The presence of tumor within pulmonary blood vessels is defined as pulmonary tumor embolism (PTE). The most common tumors associated with this disease are renal cell carcinoma and hepatocellular carcinoma. Patients suffering from pulmonary tumor embolism present with progressive, unexplained dyspnea [1,2], usually initially attributed to other causes. Affected patients may or may not have an established diagnosis of underlying malignancy. Diagnosis is usually established post mortem [4].

Case Presentation: A 52-year-old woman with no significant medical history presented with 3 days of chest discomfort, cough and shortness of breath. Upon presentation she was noted to be in respiratory distress. Other than her distress, there were no red flags in either her history or physical examination and nothing to further focalize the cause. Her laboratory findings revealed a troponin of 0.153 ng/mL and d-dimer of 4.8 μg/mL. Electrocardiogram revealed sinus tachycardia of 130 beats per minute without other abnormalities. A computed tomography angiography (CTA) of the chest was performed which was negative for pulmonary embolism or other obvious intra-thoracic process. She was admitted to the hospital for ongoing treatment and assessment for hypoxia out of proportion to findings. Her admitting diagnosis was non-ST elevation myocardial infarction (nSTEMI) and she was started on an intravenous heparin drip. During the hospital stay, patient’s oxygen requirement kept increasing requiring her to be upgraded to Medical Intensive Care Unit (MICU). An echocardiogram was performed which showed septal flattening during systole and diastole, consistent with right ventricle volume overload. These findings increased suspicion for pulmonary embolism or other obstructive process. Two hours after transfer to the MICU, she suffered a cardiac arrest. Despite Advanced Cardiovascular Life Support, return of spontaneous circulation was not obtained. Autopsy revealed a 9 cm Cholangiocarcinoma with significant intravascular permeation by the mass. The vascular involvement of the tumor had resulted in extensive tumor embolization of small arterial branches of the pulmonary arteries.

Discussion: Patients presenting with significant hypoxic respiratory distress are typically suspected to have an intra-parenchymal lung process or pulmonary embolism. Cardiac etiologies are typically next in line in the differential diagnosis. Pulmonary tumor embolism, however, is usually suspected in patients with known malignancy. When a patient presents with sudden onset of shortness of breath and no known malignancy, a diagnosis of PTE is hard to make; in younger patients, this is even less often suspected.

The patient in this case had no known malignancy and CTA of the chest was performed which was negative for pulmonary embolism. One needs to have high level of suspicion to establish a diagnosis of PTE as traditional test such as CTA can be negative and novel diagnostic strategies such as pulmonary artery catheter aspiration, transbronchial biopsy or surgical lung biopsy needs to be considered for diagnosis.

References

NEW JERSEY CLINICAL VIGNETTE POSTER FINALIST - ALEXANDRE SHEHATA, MD

The Chase for a Diagnosis: CNS Toxoplasmosis or Cerebral Lymphoma in 2018, a Challenging Case in Progress.

Authors: Alexandre Shehata MD, Paul Wojcieszek MD, Ashraf Elamin MD, Aquino Williams BS, James Brooks BA, and Isaac Soliman MD

Introduction: Central Nervous System (CNS) Toxoplasmosis, a well-documented opportunistic infection closely tied to Acquired Immune Deficiency Syndrome (AIDs) has seen a gradual decline in incidence since the advent of highly active anti-retroviral therapy (HAART). Commonly known to masquerade as CNS lymphoma, a proper diagnosis and early treatment is of essence. Unfortunately, many of the problems faced in the 1980s in regards to procuring an accurate diagnosis and treating CNS toxoplasmosis seem to still be relevant forty years later.

Case Presentation: We present a case of a 32 year old female with no reported medical history who presented with worsening fatigue, weight loss, urinary and fecal incontinence. She had previously been hospitalized at another facility for three weeks with no reported diagnosis upon discharge. On presentation the patient was bradycardic and hypotensive despite fluid resuscitation. Laboratory results revealed positive Human Immunodeficiency Virus (HIV) Ag/Ab combination, CD4 count of 28, and viral load of 380,000. The patient later attested to acknowledging her HIV diagnosis six years prior, but never sought treatment until one year ago. At that point in time, she took elvitegravir/cobicistat/emtricitabine/tenofovir as HAART for six month however discontinued therapy due to side-effects of lethargy and hair loss. On admission, magnetic resonance imaging (MRI) revealed ring-enhancing masses extending across the genu of the corpus callosum consistent with a neoplastic or infectious process. Treatment for presumed toxoplasmosis was started with sulfadiazine and pyrimethamine with leucovorin rescue therapy with the addition of steroid pulse therapy for a week. HAART was deferred for two weeks considering her risk of developing Immune reconstitution inflammatory syndrome (IRIS). Repeat MRI was scheduled per Infectious Disease Society of America’s recommendations in two weeks.

Toxoplasma Immunoglobulin G titers resulted positive, although Immunoglobulin M titers were negative. Patient continued to decline with frequent febrile states, oral candidiasis, nonsustained ventricular tachycardia, transaminitis, worsening myopathy and septic shock. Two weeks later, despite some improvement in her initial presenting symptoms, repeat brain MRI showed no significant change. Concern for a lymphoma remained. Nuclear magnetic resonance spectrometry with thallium uptake was negative. HAART was initiated at this juncture, under the cover of the anti-toxoplasmosis treatment. A repeat brain MRI two weeks later remained unchanged, prompting the decision for brain biopsy, which confirmed the presence of toxoplasms on immunohistochemistry with no neoplastic cells. She was discharged on Clindamycin-Pyrimethamine-Sulfadiazine-Folinic Acid with further imaging scheduled in six weeks which - to our dismay- showed worsening of the lesions.

Discussion: Despite the patients’ compliance with treatment and the appropriate medical management, the patient has yet to show any significant improvement in her condition, begging the question: “could a lymphoma be present despite negative tests and biopsy? Or have the forty-year old anti-toxoplasma medications become ineffective in treating CNS toxoplasmosis in 2018?”

References

Acute Fulminant Hepatotoxicity after a single dose of PTU: An Unusual Case

Authors: Stephanie Vuong, DO, Sonal Pathak, MD

Introduction: PTU induced hepatotoxicity is a well known potential complication. Onset is typically 2-12 weeks after initiation and is generally short-lived with discontinuation. We present a patient with an impending thyroid storm who developed fulminant hepatotoxicity 9 hours after a single dose of PTU.

Case Presentation: A 38 year old woman with PMH of multinodular goiter, hyperthyroidism, and iodine allergy presented with palpitations, chest discomfort, and dyspnea. She had been non-compliant with methimazole for the last 2-3 years and was recently restarted on 10mg daily. In the ED she was in atrial fibrillation with ventricular rate of 170 bpm and an LVEF of 20%. Labs revealed a suppressed TSH (0.01 mIU/L), elevated free T4 (3.48 ng/dL), elevated free T3 (9.90 pg/mL), normal liver transaminases, and a Burch Wartofsky score of 45. 9 hours after treatment with propranolol and PTU 200mg her AST and ALT levels peaked at 1127 IU/L and 1004 IU/L, respectively. PTU was stopped after the first dose, followed by failure of a dose taper of methimazole due to persistently worsening LFTs. Solumedrol was initiated with improvement in both LFTs and thyroid function tests allowing for reintroduction of methimazole without adverse effects. On discharge, liver enzymes normalized to AST 38 IU/L and ALT 163 IU/L with an LVEF of 40%. The patient has since had progressive increases in methimazole doses up to 40mg daily on an outpatient basis without hepatotoxicity. A 1 month follow up showed complete normalization of liver enzymes.

Discussion: Incidence of PTU induced liver injury is estimated to be at least 1 per 1000 persons with acute liver failure occurring in 1 in 10,000 adults. Timing of the injury post PTU exposure has not been well studied, however general consensus is about 2-12 weeks after drug initiation with resolution typically occurring after drug cessation. In our case, fulminant hepatotoxicity was induced by one dose of PTU in the setting of an impending thyroid storm with reduced EF. The rapidity in which this occurred was unexpected as autoimmune and viral hepatitis tests were negative. It is unclear whether fulminant hepatotoxicity was precipitated primarily by PTU or a reduced EF, which also played a role. It is important to be mindful of complications that can occur from thioamides, particularly PTU. Exercise caution as well as be hypervigilant when using this medication over methimazole.
NEW MEXICO CLINICAL VIGNETTE POSTER FINALIST - REEM ALKILANY, MD

A puzzling skin lesion

Authors: Reem Alkilany, MD, Linda Macdonald, MD

Introduction: Livedoid vasculopathy (LV) is a chronic, non-inflammatory, vaso-occlusive condition of cutaneous blood vessels. It can either be primary (idiopathic) or secondary associated with coagulation abnormalities or fibrinolysis. LV incidence is 1:100,000 with a female to male ratio of 3:1. It presents initially as telangiectatic, painful purpuric papules and plaques evolving to an ulcerative stage followed by star-shaped porcelain-white atrophic plaques, Atrophie Blanche (AB), with peripheral telangiectasia and hyperpigmentation. Here we present a case of a sudden onset medial malleolar skin ulcer resistant to antibiotics but improved with aspirin therapy.

Case Presentation: A 62-year-old male with past medical history of DM2, HTN, dyslipidemia, obesity, iron deficiency, OSA, and asthma presented with a right medial malleolar shallow ulcer without significant surrounding skin changes that had rapidly evolved from a small pimple. The patient was otherwise asymptomatic and denied previous similar symptoms, trauma, recent medication changes, hiking, traveling or insect bite. He denied personal or family history of bleeding or clotting disorders or autoimmune diseases. Vital signs were stable, laboratory workup was significant for a mild leukocytosis, with WBC of 12,600 with a normal differential, and plasma glucose of 209mg/dL. Wound fluid sent for C&S was positive for MSSA and S. alpha hemolyticus. The patient was started on clindamycin 300mg q6h, yet his ulcer continued to progress in size and thickness and developed rolled edges with surrounding erythema. Despite multiple debridements and continuing antibiotic therapy, the entire wound became friable with areas of necrosis and moderate serosanguinous to bloody drainage but no purulence or exposed tendons. Physical features of the lesion and the initial biopsy findings raised concern for pyoderma gangrenosum, so the patient was started on cyclosporine100mg TID and methylprednisolone 4mg daily. Despite this approach, the ulcer continued to enlarge and pain continued to worsen, limiting the patient’s mobility. This mandated further workup including evaluation for autoimmune and coagulation disorders with negative results. The ulcer tissue biopsy was later evaluated by a dermatopathologist and a diagnosis of livedoid vasculopathy was made. The patient was started on aspirin 325mg daily, and cyclosporine and methylprednisolone were discontinued. With continued wound care and debridement there was a notable decrease in ulcer size, resolution of serosanguinous drainage, and significant improvement in pain.

Discussion: Treatment for LV requires multiple approaches, most importantly pain management with acetaminophen, NSAIDS or gabapentin. Treatment should also include wound care, maintaining a clean, moist environment, limiting thrombosis risk with antiplatelet, antithrombotic, fibrinolytic, and/or anticoagulant medications, and environmental modification such as smoking cessation. This rare disorder can be confused with other etiologies such as pyoderma gangrenosum or vasculitis. Inappropriate diagnosis leads to inappropriate management with antibiotics and steroids resulting in disease progression, increased pain and impaired quality of life which can be avoided by early recognition.
Hemophagocytic Lymphohistiocytosis due to CMV infection

Authors: Amir Anabtawi MD, Reem Alkilany MD, Jacklyn Nemunaitis MD, Mary Lacy MD

Introduction: Hemophagocytic lymphohistiocytosis (HLH) is a rare disease characterized by an overwhelming immune system activation and subsequent heightened inflammation state. In this case report, we report a case of CMV-triggered HLH. We also highlight the importance of early detection and the significance of trigger identification in the treatment of HLH.

Case Presentation: A 27-year-old man with a past medical history of alcohol use disorder and newly diagnosed, untreated HIV who was transferred to our hospital from an outside facility for the work-up of persistent fevers. Upon presentation, he reported fevers, chills, night sweats and abdominal pain. He denied nausea, vomiting, diarrhea, or constipation. He denied a personal or family history of recurrent infections, blood disorders, malignancy, or autoimmune disease. Vital signs were significant for a temperature of 38.1°C and heart rate of 102 bpm. Examination was notable for cachexia, oral thrush, mild hepatomegaly, and muscular weakness. Laboratory workup: Pancytopenia Critically low CD4 count of 3 cells/uL. CMV viremia with more than 10 million copies/mL. Other infections including EBV, HSV, TB, PJP were negative. Abdominal ultrasound revealed mild hepatic steatosis with hepatomegaly but no splenomegaly. Hemophagocytic lymphohistiocytosis (HLH) was diagnosed based on the 2004 diagnostic criteria. CMV was identified as the most likely trigger for his secondary HLH. He was started on prednisone as well as valganciclovir. Within 2 weeks, the patient began to improve, he was afebrile, his bone marrow function started to recover, and his ferritin levels dropped from 15,462ng/mL to 7,930ng/mL. The patient was started on anti-retroviral therapy prior to discharge.

Discussion:

- HLH is a rare disease caused by a dysfunction of cytotoxic T cells and NK cells. This T cell/NK cell dysregulation causes an aberrant cytokine release, resulting in proliferation/activation of histiocytes with subsequent hemophagocytosis.
- HLH can be either primary, i.e. due to an underlying genetic defect or secondary, associated with malignancies, autoimmune diseases or infections.
- Infectious triggers are most commonly due to viral infections mainly of the herpes group, with Epstein-Barr Virus being the most common cause. [2]
- HLH commonly presents with vague symptoms of systemic illness such as fever, weakness and fatigue.
- In the case we report, the patient presented with unexplained fever and was found to have HLH due to CMV infection in the setting of HIV infection. Interestingly, treatment of CMV infection led to remission of HLH.
- In a retrospective study of HLH disease in the setting of HIV infection, there were only 6 cases of CMV-triggered HLH. Moreover, all of them had other co-infection with mycobacterial infection. [3]
- Untreated HLH carries a high mortality rate. Thus, prompt diagnosis and correct identification of the trigger is imperative to management. Our patient had significant improvement after starting steroid therapy along with treatment for CMV.

References


NEW YORK CLINICAL VIGNETTE POSTER FINALIST - NICHOLAS BECCARINO, MD

Myocardial Infarction as the Initial Manifestation of Systemic Lupus Erythematosus in a 32-Year-Old Woman

Authors: Nicholas J. Beccarino, MD, David Altszuler, MD, Scott Butler, MD, Sohah N. Iqbal, MD

Introduction: Systemic lupus erythematosus (SLE) is a heterogeneous syndrome leading to significant multiorgan pathology. Subclinical inflammation in patients with undiagnosed SLE plays a significant role in coronary disease and can present among patients without evidence of active inflammation. We present a case of a 32-year-old female presenting with VF arrest in the setting of a STEMI as the first manifestation of undiagnosed SLE.

Case Presentation: A 32-year-old female was transferred to our institution after a cardiac arrest. The patient was well until 4 months prior when she developed exertional chest pain, ultimately discharged from an outside hospital with presumed myo-pericarditis in the setting of a mildly positive troponin. On the morning of presentation, the patient awoke with chest pressure before collapsing in front of her husband who drove her to the nearest hospital. She arrived pulseless in a wide complex rhythm with return of spontaneous circulation 37 minutes after her collapse. Electrocardiogram was notable for ST elevation in V4-6. Subsequent cardiac catheterization demonstrated severe proximal LAD pathology and an occluded diagonal consistent with atherosclerosis and deemed suitable for coronary artery bypass surgery. The patient underwent 24 hours of hypothermia with full neurological recovery after rewarming. The patient had no clear coronary risk factors but following a thorough workup was diagnosed with SLE in the setting of lymphopenia, positive anti-nuclear, anti-DsDNA, and anti-Smith antibodies. The patient underwent quadruple coronary bypass and was discharged home on post-operative day 5.

Discussion: Cardiovascular pathology is the leading cause of mortality in patients with SLE, particularly among young women who experience a relative risk of myocardial infarction fifty times that of those without SLE.1 Accelerated atherosclerosis as well as spontaneous coronary artery thrombosis, aneurysm and dissection, and vasculitis have all been reported in asymptomatic patients subsequently found to have SLE.2-6 While traditional cardiac risk factors play a role, multiple studies have shown that they do not fully account for increased cardiac risk.7 Research into the interaction between inflammatory cells and the underlying atheroma suggests that chronic inflammation contributes to plaque accumulation and rupture. Endothelial dysfunction and integration of inflammatory cells into areas of early endothelial damage and resulting cytokine release has been shown to increase lipid uptake and plaque formation.8,9 Furthermore, macrophages and T lymphocytes are often present at the site of plaque rupture, suggesting that inflammation ultimately plays a role in acute coronary syndromes.10 This patient’s presentation was the result of premature atherosclerosis accelerated by a chronic inflammatory state in the setting of subclinical SLE. We believe that the threshold for coronary evaluation should be low among patients with SLE, and that SLE should be considered in the workup of any young female with angina or a cardiomyopathy, even in the absence of symptoms, to prevent the potentially fatal complications of an untreated inflammatory state.

References

An Unusual Presentation of Carbapenem Neurotoxicity

Authors: Carlos Ceron, MD, Rafael Valle-Salinas, MD, Nashreen Anderson, DO, Francis O’Neil, MD, Dipti Kothari, MD

Introduction: Ertapenem, a novel broad-spectrum antibiotic, has been primarily used for the treatment of complicated aerobic and anaerobic gram-positive and gram-negative bacteremia. Drug-induced neurotoxicity is an uncommon adverse effect that has been previously reported in the literature.

Case Presentation: A 53-year-old male with a past medical history significant for end-stage renal disease (ESRD) on hemodialysis, diabetes mellitus type 2, and hypertension, presents to the hospital complaining of shortness of breath and foot pain for 1 week. Prior to admission, patient was admitted to another facility and was treated for left foot osteomyelitis with a six-week course of intravenous (IV) antibiotics. He also had a surgical amputation of the left foot 5th metatarsal. Physical examination was significant for bibasilar crackles and left foot erythema with profound edema. MRI performed revealed osteomyelitis of the 5th metatarsal base, with 4th metatarsal involvement. Wound cultures of the left foot were obtained, and patient was started on broad-spectrum IV antibiotics. Few days later, cultures showed positive polymicrobial growth, significant for ESBL producing Enterobacter cloacae. Upon reported final antibiotic susceptibility results, guided therapy with Ertapenem was initiated. Dosage and frequency were adjusted, in the setting of ESRD on hemodialysis. Medical management was continued, along with hemodialysis sessions every 48 hours. After 28 days of antibiotic therapy, patient was found to have new onset dysarthria, with no other focal neurological deficits. CT and MRI studies of the head/brain showed no acute intracranial pathology that would explain the patient’s isolated dysarthric speech. Over the next 3 days, patient continued with unresolved dysarthria, along with new onset neuropsychiatric manifestations, which included visual and auditory hallucinations. EEG performed revealed no spike wave abnormalities that would suggest epileptic activity. Further neurological testing was completely negative. Given the rare, but potential neurotoxic effect of Ertapenem, it was decided to discontinue therapy. Patient was started on Tygecycline and TMP-SMX. Exactly 96 hours of discontinuation of Ertapenem, patient’s neurological manifestations showed complete resolution, with no residual deficit. Over the course of the remaining hospital stay, patient had no recurrence of neurological symptoms and successfully completed a total of 42 days of IV antibiotic therapy. In view of significant clinical improvement and completion of therapy, patient was discharged from the hospital.

Discussion: The prevalence of seizure activity secondary to Ertapenem is estimated to be less than 1%, while non-seizure neurotoxicity is even rarer those symptoms occur due to a structural similarity and antagonism with GABA receptors, but also N-methyl-D-aspartate (NMDA) receptors.

Our patient presented with an uncommon adverse effect of carbapenems with new onset of neuropsychiatric symptoms and negative neurological investigation for other possible etiologies as metabolic encephalopathy, central nervous system infection or cerebrovascular accidents with an evident clinical improvement after discontinuation of ertapenem.

References

A Rare Case of Gastric Cancer Metastasizing to the Colon

Authors: Bing Chen, MD, Mount Sinai St Luke’s and West, New York, Neelesh Rastogi, MD, Mount Sinai Beth Israel, St Luke’s, and West, New York, Shu Min Lao, MS4, NYIT College of Osteopathic Medicine, New York

Introduction: Gastric cancer is the sixth most common cancer and third leading cause of cancer death worldwide. Patients may present with distant metastatic disease. However, metastases rarely involve the colon. We report a case of severe iron deficiency anemia with symptoms of right lower abdominal pain and intermittent constipation found to have primary gastric signet-ring cell carcinoma with a solitary colonic metastasis.

Case Presentation: A 39-year-old African American male with iron deficiency anemia was referred to the emergency department by his primary care provider for a decreased hemoglobin level of 7.1 (baseline of 11.2 six months prior). He complained of mild postprandial right lower quadrant abdominal pain associated with intermittent constipation for a few days prior to his admission without bloody stool or melena. On examination, he was hemodynamically stable and noted to have mild tenderness and firmness in the right lower quadrant of the abdomen. CT abdomen and pelvis with contrast showed extensive soft tissue thickening of the ascending colon, peritoneal cavity, and gastric wall, as well as extensive enlarged nodes in the upper abdomen. Upper endoscopy showed friable mucosa with thickened folds in the proximal stomach suggesting an infiltrative process. Colonoscopy revealed a malignant-appearing, intrinsic severe stenosis measuring 1cm (in length) x 1cm (inner diameter) in the distal ascending colon. Biopsies from both sites were obtained. Stomach biopsy confirmed poorly differentiated adenocarcinoma, diffuse type with signet ring cells. Biopsy of the abnormal colonic mucosa surrounding the stenosis revealed poorly differentiated adenocarcinoma histologically consistent with metastasis from gastric primary, which was confirmed by mucicarmine and pancytokeratin stains. Diagnostic laparoscopy with peritoneal implant biopsy was also consistent with metastatic adenocarcinoma. Immunostain E-CAD is positive, Her2 is equivocal (2+). CT Chest with contrast showed calcified left lower lobe nodules and thoracic lymph node metastases. The patient was diagnosed with stage IV gastric adenocarcinoma and is currently enrolled in a clinical trial.

Discussion: The most common metastatic distribution of gastric cancer is the liver, peritoneal surfaces, and nonregional or distant lymph nodes. Colonic metastases from gastric cancer are rare, typically attributed to peritoneal carcinomatosis, linitis plastica, and poorly differentiated adenocarcinoma [1]. The most likely method of metastasis in this case could be via peritoneal seeding. Several cases of multiple colonic metastases from poorly differentiated gastric adenocarcinoma, presenting as colonic polyposis or multiple flat elevated lesions, have been reported [2]. We demonstrated a very rare case of primary gastric signet-ring cell carcinoma with a solitary colonic metastasis, presenting as severe stenosis.

References

NEW YORK CLINICAL VIGNETTE POSTER FINALIST - PATRICIA DE LIMA, MD

A Case of Metformin Toxicity Resulting in Vasoplegic Shock

Authors: Patricia De Lima, MD, Samuel Rednor, DO, Manoj Karwa, MD., AECOM, Montefiore Medical Center, Wakefield Division.

Introduction: Although metformin is well tolerated and carries a mostly benign side effect profile, metformin-associated lactic acidosis (MALA) is a severe and potentially fatal reaction to the drug, particularly in patients with concomitant tissue hypoperfusion. Albeit rare, MALA is associated with a high case fatality rate due to vasopressor-refractory shock, or vasoplegia. It is therefore imperative to recognize that an acute process, such as dehydration, may drastically increase the likelihood of developing this life-threatening condition in an otherwise metformin-suitable diabetic. Here we present a case of MALA-induced vasoplegia that was successfully treated with methylene blue (MB) as rescue therapy when conventional management had failed.

Case Presentation: A 64-year-old man with DM (metformin) and chronic kidney disease (losartan) presented with four days of abdominal pain, vomiting, and intolerant of anything other than minimal liquids. On presentation, he was afebrile, hypertensive (199/135mmHg), tachycardic (111/minute), tachypneic (24/minute), with a normal pulse oximetry on ambient air. His abdomen was diffusely tender but soft and non-distended. Labs were remarkable for WBC 17.6, pH 6.87, lactate 16, BUN/Cr 105/12.28, and bicarbonate <5. Bedside ultrasonography revealed a hyperdynamic left ventricle and no free intra-abdominal fluid. Semi-upright abdominal X-ray was significant for mildly dilated small bowel loops. Patient became critically hypotensive, was emergently intubated, started on broad-spectrum antibiotics, stress-dose steroids, and intermittent hemodialysis (IHD). Surgery was consulted, and while unable to exclude abdominal catastrophe as inciting event, there was no role for emergent exploratory laparotomy given his acute hemodynamic instability. Shock was ultimately refractory to four vasopressors and clinical deterioration progressed notwithstanding IHD and optimizing ventilation (nebulized prostacyclin and pronation). At this juncture, he was transferred to neighboring facility for continuous venovenous hemodialysis (CVVHD) and possible extracorporeal membrane oxygenation (ECMO).

There, the patient underwent CVVHD and was administered a 100mg (1.5mg/kg) intravenous bolus of methylene blue (MB) as rescue therapy. His vasopressor requirements diminished precipitously - within 6 hours, his total requirement had been halved. Within 24 hours of MB, lactate normalized. On hospital day four, the patient was successfully extubated and resumed on IHD. He was discharged to acute rehabilitation on hospital day 14. Notably, the septic work-up was negative and the metformin level, sent on presentation, was 18mcg/mL [1-2mcg/mL].

Discussion: Although MB is used in various clinical contexts (post-cardiac surgery and in various toxicology settings), further research into its use as adjunct therapy in refractory shock is prudent. Specific to this case, MB acts by inhibiting guanylate cyclase while simultaneously scavenging nitric oxide and inhibiting its synthesis. This counteracts the shock-inducing activation of endothelial nitric oxide synthase by metformin. Given the ubiquity of DM in the US and the role of metformin in its treatment, clinicians must be prepared to identify and treat MALA-induced vasoplegia – the consequences of not doing so may prove catastrophic.
Avoiding “Rash” Conclusions: A case of adult onset IgA vasculitis with cutaneous, gastrointestinal, and renal involvement

Authors: Amanda Dowden, MD; William West, MD

Introduction: Henoch–Schönlein purpura (HSP) is a systemic vasculitis with prominent skin findings. Because of the morbidity associated with the renal complications of HSP, it is important to consider when thinking about leukocytoclastic vasculitides.

Case Presentation: A 59 yo Asian man with history of psoriasis presented with 2 weeks of abdominal pain, decreased PO intake, and a progressively worsening rash which had started on his legs, and since spread to his arms, hands, and palms. Initial labs revealed a WBC of 27k with bands and a leukocytic predominance and no eos, plt of 301, Cr of 2.0, ESR of 96 mm/hr, and CRP > 270 mg/L. Physical exam was notable for scattered non-blanching petechiae on his arms, and palpable purpura with ulceration on both legs. CT A/P showed proximal small bowel dilatation with wall thickening and mesenteric inflammation. Skin biopsy revealed pustular vasculitis positive for IgA. These findings were consistent with systemic IgA vasculitis (HSP) with cutaneous and GI involvement. He was treated with pulse steroids and his GI symptoms and rash rapidly improved. Despite the treatment, he then developed hematuria concerning for renal involvement. His course was also complicated by a SMV thrombus. A hypercoagulable workup was unrevealing. He was treated with a prednisone taper, started on long-term anticoagulation, and was discharged home.

Discussion: Despite being the most common vasculitis in children, HSP is fairly uncommon in adults, with about 3 - 14 cases per million. Renal involvement tends to be more severe in adults, with higher reports of nephrotic syndrome, hypertension, and elevated creatine. While the GI tract is often involved in adults, severe manifestations such as intussusceptions, massive bleeding, and perforation, are much less common. Typical EGD findings include erythema, swelling, and submucosal hemorrhage.

HSP tends to be self-limiting and the mainstay of treatment is supportive care. However, early recognition and treatment is important in adults to prevent complications. Steroids should be considered in severe cases, though their use is controversial as the data is limited and literature has shown that they do not actually prevent the onset of renal or GI disease (bearing in mind that a majority of the data is from the pediatric population). Regardless, by reducing the edema in the intestinal wall, steroids are effective in treating abdominal pain associated with HSP.

HSP is a proinflammatory state, with elevated levels of TNF-α, fibrinogen, and IL-6. Patients also have impaired activation of fibrinolysis (7). Despite this fact, thrombotic events are a rare complication. While prior case reports show that HSP-associated thrombus was associated with higher levels of plasma factor VIII and homocysteine prothrombotic disease states, such as antiphospholipid antibody syndrome, our patient’s hypercoagulable work-up was unremarkable.

References

Acute reversible chorea secondary to vitamin B12 deficiency

Authors: Mohammad I. Ghanbar, Olufunke G. Adeusi, Radfan Gazali, Department of Internal Medicine, Icahn School of Medicine, Mount Sinai St Luke’s-West Hospital, New York NY.

Introduction: Vitamin B12 deficiency has been linked to several neuropsychiatric manifestations including reversible movement disorders. Whereas movement disorders secondary to depleted Vitamin B12 has been well documented in the pediatric population; very few reports of its occurrence have been recognized in adults. We present a case below of an elderly woman found to have chorea attributed to low levels of vitamin B12.

Case Presentation: A 79-year-old right-handed woman with a history of hypertension was admitted with a two-week history of progressively worsening choreiform movements of the limbs and head. The patient’s daughter noticed the movements to be more prominent in the upper compared to the lower limbs. Review of the systems was only significant for mild memory impairment in the weeks prior to symptom onset. History was negative for any alcohol consumption. Her vital signs were normal. Physical Examination revealed her to be oriented to place and person, with slow speech. Neurological exam was significant for frequent choreiform movements of the face, upper and lower extremities, with hyperreflexia in all extremities. Plantar reflexes were down-going, with intact sensation and motor strength. Her work up included a complete blood count which showed an elevated mean corpuscle volume of 113 fl with a hemoglobin level of 12.0 g/dL, undetectable Vitamin B12, elevated homocysteine of 45.4 umol/L (reference <11 umol/L) and methylmalonic acid levels of 1.83 umol/L (reference <0.4 umol/L). Her thyroid stimulating hormone, syphilis and peripheral smear testing were normal. A paraneoplastic antibody panel, and pernicious anemia antibodies were negative. Magnetic resonance imaging of the brain showed only chronic small vessel ischemia. EEG testing did not reveal any epileptiform activity. The patient was treated with parenteral cyanocobalamin with concurrent oral replacement. She improved rapidly with resolution of her chorea prior to discharge. At 1-week follow-up the patient remained symptom free and had returned to baseline activity.

Discussion: Vitamin B12 deficiency is known to cause a number of neurological abnormalities, most notably subacute degeneration of the cord and reversible dementia. Movement disorders secondary to its low levels in adults have been described in only a few case reports. The explanation of such phenomenon is thought to be secondary to the elevation of homocysteine due to the lack of Vitamin B12. Homocysteine accumulation in the brain stimulates the N-methyl-D-aspartate receptors leading to excitation of the basal ganglia and a consequent choreiform movement disorder. Methyl tetrahydrofolate elevation due to Vitamin B12 deficiency has been proposed to also play a role by invoking additional stimulation of the kainic acid receptors; which have shown to induce Huntington’s like symptoms in animal models. This case highlights the importance of testing for vitamin B12 in patients presenting with new onset choreiform movements. Correction of the deficiency leads to the resolution of symptoms.

References

NEW YORK CLINICAL VIGNETTE POSTER FINALIST - NIYATI GORADIA, MD

THE USE OF ROMIPLOSTIM FOR THE TREATMENT OF SEVERE REFRACTORY THROMBOCYTOPENIA ASSOCIATED WITH EVANS SYNDROME.

Authors: Niyati Goradia, MD1; Lewis Steinberg, MD1, 1. Jacobi Medical Center/Albert Einstein College of Medicine, Bronx, NY 10461

Introduction: Evans syndrome is an uncommon condition due to the occurrence of two or more hematologic cytopenias, most likely autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP). First line therapy is often corticosteroids and or intravenous immunoglobulin (IVIG) to which most patients respond. We present a case of Evans syndrome which was unresponsive to multiple lines of treatment including splenectomy and IVIG, who responded to Romiplostim with a normalization of platelet count.

Case Presentation: A 59-year-old male from Albania with a past medical history of ITP presented with a feeling of weakness and shortness of breath to our hospital. His ITP had been treated in the prior month with a Rituximab infusion and he was currently on a tapering dose of corticosteroids.

The initial laboratory workup showed anemia (9.2 gr/dl), severe thrombocytopenia (14,000/mm3), elevated lactate dehydrogenase (769 units/L) and an elevated total bilirubin (4mg/dl), Coombs test was positive and peripheral smear showed microcytic hypochromia with normal appearing white blood cells, and very low platelets and no schistocytes. The patient was thus diagnosed with Evans syndrome. He was initially treated with steroids and intravenous immune globulin (IVIG), however, his platelets did not show any significant response. He then underwent splenectomy as a second line treatment. However, his thrombocytopenia worsened after surgery (8,000/mm3). He was then given four infusions of Rituximab, his platelets still continued to fall. Apart from this, the patient was also found to be actively hemolyzing as evidenced by his consistently low haptoglobin, rising LDH and continued reticulocytosis even after the above mentioned treatments. He was given a trial Romiplostim on day 21 of his hospital admission. The patient had a remarkable response, with his platelets rising from 8 to 241 within 4 days of receiving Romiplostim. He was also subsequently started on Mycophenolate for treating his immune-mediated hemolysis.

The patient only required 1 dose of Romiplostim and his platelets have been stable since. He was subsequently discharged and followed in hematology clinic 4 weeks after and continued to have a stable platelet count.

Discussion: Romiplostim is a thrombopoietin(TPO) receptor agonist that acts by stimulating the production of megakaryocytes and ultimately platelets in the bone marrow by binding and activating the TPO receptor. Its use in Evans syndrome has been reported in a few prior case reports as a bridge to splenectomy. The development of thrombocytopenia in Evans syndrome may be associated with insufficient platelet production as well as increased autoimmune destruction. While corticosteroids and other immune modulating agents may decrease platelet destruction, TPO agonists aid in increasing platelet production.
Pulmonary Tuberculosis Has Gut Feeling: A Case Of Pulmonary Tuberculosis Presenting With Gastrointestinal Symptoms

Authors: Kyaw Htun (M.B;B.S); Kapilkumar Manvar (MD); Chandrika Chandrappa (MD)

Introduction: Tuberculosis (TB) is relatively rare in developed countries and intestinal TB is rarer still, comprising only about 1-3% of TB cases. Approximately, 20% of patients with intestinal TB have co-existing pulmonary TB. We present a case of active pulmonary TB with diffuse colonic involvement manifesting predominantly with gastrointestinal (GI) symptoms.

Case Presentation: A 42-year-old male immigrant, without significant past medical history, presented with a 3-month history of right lower abdominal pain, watery diarrhea and weight loss. He reported episodic fever with chills. He had prior hospitalization for similar symptoms, treated with antibiotics without substantial improvement. The patient had right lower quadrant abdominal tenderness without guarding. Leukocyte count was 14,300 with lymphocytes of 13%. He was admitted with a preliminary diagnosis of inflammatory bowel disease (IBD). CT abdomen showed right mid-abdominal small bowel intussusception with terminal ileitis and nonspecific colitis of ascending colon. CT chest revealed bronchiectasis with left apical consolidation. Colonoscopy showed scattered sessile polypoid vs pseudopolypoid lesions with inflammation in sigmoid, descending and ascending colon, and terminal ileum. Biopsy showed cryptitis, crypt abscesses and extensive necrosis. Necrotic granuloma with multi-nucleated giant cells and acid-fast bacilli (AFB) were identified. Sputum smear and cultures were positive for Mycobacterium Tuberculosis (MTB). HIV was negative. Diagnosis was confirmed as active pulmonary TB with diffuse colonic involvement. 4-drug regimen was started, and a month later, the patient was doing well with no residual symptoms. Surveillance of close contacts revealed one child with active pulmonary TB and another with positive QuantiFERON.

Discussion: TB remains a healthcare problem in the U.S. Intestinal TB is usually caused by MTB and rarely by Mycobacterium Bovis. GI infection occurs by ingestion of infected sputum, hematogenous and lymphatic spread from pulmonary foci, and contiguous spread from adjacent organs. The most common symptoms are abdominal pain, weight loss, fever and diarrhea. These non-specific symptoms frequently result in a misdiagnosis of other infections, IBD or tumor. Once suspected, the mainstay of diagnosis is colonoscopy. Ileocecal region is the most common site of involvement. Colon may be segmentally involved but diffuse colitis is rare. There may be ulcers, nodules, deformed ileocecal valve and strictures. Polypoid lesions seen in our patient are rare findings. Granulomas are found in up to 70% whereas caseating necrosis and AFB are identified only in 10-20% of cases. Culture can be negative in up to 70%. In view of its non-specific presentation, intestinal TB in the absence of concurrent pulmonary involvement poses a diagnostic challenge. In our case, previous workup in earlier admission missed the diagnosis because of the non-specificity of the patient’s symptoms. A high index of suspicion for intestinal TB should be maintained whenever the patient’s socio-economic background and clinical features suggest the diagnosis.

References

Case of fatal Acute Pulmonary Blastomycosis presenting as ARDS in immunocompetent grain inspector

Authors: Muhammad Idrees, Crischelle Magaspi, Deborah Sentochnik

Introduction: Blastomycosis is an infection caused by fungus Blastomyces. It is endemic in Ohio-Mississippi river valley and upstate New York. We present the case of severe pulmonary Blastomycosis in a middle aged immunocompetent grain inspector.

Case Presentation: 55 year old male with no significant past medical history who works as imported organic grain inspector for government presented to us with two day history of sudden onset shortness of breath, dry cough and high grade fever. Patient recently had inspected the wheat imported from Africa, since then he started having symptoms. CXR was concerning for multifocal consolidation. White count was 22000, lactic acid was normal. Patient was initially thought to have community acquired pneumonia and was started on IV antibiotics. However he declined over next few days day and developed severe acute hypoxic respiratory failure requiring intubation.

CT angiography was negative for pulmonary embolism. Blood cultures were negative. Urine strep and pneumococcus were negative. Due to concern for possible autoimmune pneumonitis he was started on IV steroid, however he declined more.

Autoimmune work up including DsDNA, C3, C4, anti-SSA, SSB, anti-u1rNP, anti-cCP, ANCA, scl-70, anti-smooth muscle ab, Polymyositis antibodies, Hypersensitivity pneumonitis panel, aldolase, TSH, HIV screen, Lyme panel, serum fungitel, respiratory viral cultures, blood cultures were negative.

Bronchoscopy with BAL was done and specimen were sent for the cytology, antigen testing and culture.

BAL aspergillus, fungitel, Flow cytometry and Cytology were negative, however BAL culture grew Blastomycosis colonies. Patient was started on Amphotericin B. Due to severe Hypoxic respiratory requiring ECMO, patient was transferred to University of Rochester where he died over next 2 days.

Discussion: Pulmonary Blastomycosis can present with wide range of symptoms and usually require high clinical suspicion for the diagnosis. Previously reports have shown that diagnosis of Pulmonary Blastomycosis have been delayed due to initial presentation with variety of symptoms which resemble other diseases. We suggest that patient from Blastomycosis endemic area should be evaluated for the possible pulmonary Blastomycosis as early intervention may be helpful.

References

An Unexpected Outcome of a Routine Preoperative Medical Consultation

Authors: Karin Karpin MD, Shitij Arora MD FACP

Introduction: The Internal Medicine Consult service is often called on for assistance with preoperative medical evaluations for patients undergoing non-emergent surgical procedures. The evaluation commonly includes perioperative mortality risk assessment and recommendations for medication adjustment. It can also serve as a safety net by identifying lethal pathology and refocusing on medical management.

Case Presentation: A 52 year-old man with uncontrolled diabetes presented with abdominal pain and inability to urinate. He was found to have Fournier’s Gangrene involving the perineum leading to an emergent surgical debridement and diverting colostomy. Three days later, the Internal Medicine Consult service was consulted to perform a preoperative medical evaluation before the patient was to be taken back to the operating room for scrotal wound closure. Physical exam was notable for tachycardia and a high output colostomy. There was no hypoxia but the patient was notably tachypneic. His potassium levels ranged from 2.5–2.9meq/L over the last five days despite repletion of nearly 120meq/day. The etiology was presumed to be gastrointestinal losses. A hypokalemia work-up was initiated by the Internal Medicine Consult service and the patient was found to have a urine potassium level less than 15mmol/L, venous blood gas with a pH of 7.505 and a pCO2 of 29.9mmHg. Serum bicarbonate was 20meq/L. This was suggestive of acute uncompensated respiratory alkalosis. A chest-x ray was normal. A contrast enhanced CT chest was obtained to look for pulmonary vascular pathology which showed a left lower lobe pulmonary embolism (PE). Surgery was deferred and the patient was started on therapeutic anticoagulation with enoxaparin with improvement in pH to 7.35 and potassium to 4meq/L after one week of treatment.

Discussion: Preoperative medical consultations are routinely performed although it remains unclear if they improve postoperative outcomes. The current literature includes studies that have found positive correlations between preoperative consultation and mortality1 as well as increased length of stay and rate of developing a complication.2 An additional study has shown that surgeons found cardiac risk stratification and optimization of cardiac medications to be the most useful aspects of the preoperative evaluation.3 This case highlights the importance of the role of the preoperative medical evaluation beyond routine cardiac risk stratification. It also highlights the importance of a systematic approach to diagnosing electrolyte disorders. We approached this patient’s hypokalemia on the basis of pathophysiology by evaluating whether decreased potassium was due to gastrointestinal losses, renal losses and/or transcellular shifts. A low urine potassium excluded renal losses. Gastrointestinal potassium losses are associated with metabolic acidosis. Acute respiratory alkalosis suggested a pulmonary pathology and since the patient had normal lung parenchyma we evaluated the pulmonary vasculature which revealed an acute PE. Hyperventilation from a PE causes a decrease in pCO2 resulting in a relative increase in HCO3- as the ratio of CO2 and HCO3- changes. This increase in HCO3- causes a transcellular shift of K+ in exchange for H+ to buffer the HCO3- resulting in hypokalemia.

References

Succumbing to the Fall: A Presentation of West Nile Virus Poliomyelitis

Authors: Akanksha Kashyap MD, Marcella Gonzalez MD, Hao Kee Ho MD, Ravi Gupta MD, Zubin Tharayil MD, Edison Gavilanes MD, Long Island Community Hospital, Patchogue, NY

Introduction: West Nile Virus (WNV) is a flavivirus transmitted via infected mosquitoes. Over 70% of WNV infections are asymptomatic; symptomatic patients often present with a self-limited flu-like syndrome with less than 1% of the infected population at risk for neuroinvasive disease. Neuroinvasive disease is classified as encephalitis, meningitis, and poliomyelitis. Poliomyelitis is a rare subtype presenting with acute and asymmetric flaccid paralysis.

Case Presentation: An 85-year-old female with no past medical history presented in the late summer after a fall due to sudden onset of generalized weakness. Prior to onset of symptoms, patient was in a good state of health and actively helped on the family farm. On initial exam, she was afebrile, with intact mentation, right facial weakness and left-sided hemiparesis. Labs were significant for rhabdomyolysis, lactic acidosis, and leukocytes on urinalysis. She was admitted and treated for a suspected cerebrovascular accident and a urinary tract infection. Brain CT and MRI were unremarkable. On day three, the patient developed a fever and weakness progressed to right-sided hemiparesis. Subsequently, her mentation declined rapidly; she was intubated and antibiotics were broadened to cover meningitis and tick-borne illnesses. Cerebrospinal fluid (CSF) analysis revealed moderate pleocytosis with increased protein and glucose. Blood cultures, Lyme titers, VZV and HIV were negative. An EEG revealed periodic lateralizing epileptiform discharges consistent with meningoencephalitis. On day nine of the hospitalization, CSF serology was positive for West Nile IgM establishing the diagnosis of West Nile meningoencephalitis with poliomyelitis. Due to her overall poor prognosis, her family withdrew care and the patient expired.

Discussion: WNV poliomyelitis is a rare, neuroinvasive disease that presents with acute and asymmetric flaccid paralysis in the presence or absence of fever or meningoencephalitis. Patients over the age of 60 with pre-existing co-morbidities with antecedent events involving outdoor activity during the summer months are at greater risk. The mortality rate of neuroinvasive disease is approximately 10% with higher rates in patients with poliomyelitis. The pathogenesis and clinical manifestations are similar to poliovirus with destruction of anterior horn cells leading to the hallmark flaccid paralysis. Diagnosis requires serologic testing for virus-specific IgM antibodies in serum or CSF via MAC-ELISA. CSF analysis generally shows lymphocytic pleocytosis with neutrophils predominating early during the illness. Routine labs and brain imaging are usually equivocal. There is no specific treatment and standard treatment remains supportive care.

Given the increasing incidence, physicians should consider WNV in their differential diagnosis of a patient who presents with asymmetric muscle weakness, particularly during the summer months. Although there is no definitive treatment, awareness of the various disease manifestations of WNV may lead to early recognition and prevent inappropriate therapy.
Creutzfeldt Jakob Disease in a Former Healthcare Worker: A Potentially Overlooked Risk Factor

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Introduction: Prion disease is a rare fatal progressive neurodegenerative spongiform encephalopathy that can be acquired, inherited, or most commonly sporadic and the most common human form being Creutzfeldt-Jakob disease (CJD). This case report explores the potential occupational risk factors of being a healthcare worker in a patient diagnosed with Creutzfeldt-Jakob disease. This raises the concern that many cases in this population diagnosed as sporadic may in truth be acquired.

Case Presentation: A 60 year old female was admitted for rapid cognitive and functional decline over 6 weeks. Three months prior, she ceased working as an OR nurse due to depression and emotional outbursts and was being treated by an outpatient psychiatrist for depression. On presentation she was aphasic, echolalic and apraxic with notable outstretched hands, clenching fists and brisk lower extremity reflexes. Her initial workup was all negative and included MRI brain, CBC, ESR, CRP, toxicology, CSF analysis, syphilis, Lyme disease, JC virus and paraneoplastic and autoimmune encephalitis. Patient was treated with benzodiazepines for catatonia, as well as an empiric treatment with steroids and IVIG for possible antibody negative autoimmune encephalitis. Despite these measures she continued to decline. A repeat brain MRI, 17 days into her hospital course showed cortical ribboning and abnormal left caudate signaling and a repeat CSF sampling was positive for RT-QuIC and 14-3-3 protein, an elevated T tau protein >4000 pg/m and an elevated neuron specific enolase of 145 ng/mL, making CJD the most likely diagnosis. Due to the known prognosis of CJD and the patient’s continued decline to the point of being nonverbal with hypertonic rigidity, startle myoclonus and concerns for aspiration she was transferred to inpatient hospice service.

Discussion: This patient lacks known established risk factors for CJD, including corneal transplants, neurosurgeries, or exposure of contaminated dura mater grafts and human pituitary growth hormones (2,3). Her history is only notable for her occupation as an OR nurse. The literature describes previous cases of physicians and nurses developing CJD but there is no conclusive evidence that being a healthcare worker is by itself a risk factor (4). It has been reported that those who work in physicians’ offices have a statistically significant greater odds of 4.6 for CJD (5). It has also been shown that prion seeding is possible on the skin and it can bind to stainless steel and surgical instruments especially if there is ineffective sterilization procedures since higher temperatures are needed to deactivate prions (6-12). As a result, we should consider the possibility of prion infectivity for both patients undergoing surgeries and healthcare workers that come in contact with surgical instruments. Consequently, we may need to re-evaluate current sterilization processes with the hope to possibly reduce the incidence of CJD thought to be sporadic in nature.

References


A unique presentation of a rare disease: Biopsy proven Systemic Lupus Erythematosus and Microscopic Polyangitis: An overlap syndrome.

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Introduction: Systemic Lupus Erythematosus (SLE) and ANCA-associated vasculitis are classically thought to be separate diseases with different pathophysiology. Rarely, the two diseases can be found together as an overlap syndrome.

Case Presentation: A 26-year-old Caucasian male with no past medical history presented with fevers, dyspnea, hemoptysis, generalized weakness, and lower extremity edema. Vitals were significant for a temperature of 103.3F, pulse of 150, blood pressure of 129/53, respiratory rate of 22 and oxygen saturation of 87% on room air. On physical exam, he was found to have moderate wheezing and rhonchi in bilateral full lung fields, moderate respiratory distress with use of accessory muscles and 2+ pitting pedal edema.

Initial blood work revealed hemoglobin of 6.4 g/dL and creatinine of 3 mg/dL. Subsequent testing revealed positive p-ANCA, MPO-ANCA, dsDNA antibody (62), ANA (1:640), with negative PR-3. CT chest revealed small bilateral pleural effusions and bibasilar ground-glass opacities. Renal CT was unremarkable. Lung biopsy showed alveolar hemorrhage without vasculitis or granulomatous inflammation. Renal biopsy revealed MPO-ANCA associated focal, necrotizing and crescentic glomerulonephritis, superimposed on class V diffuse membranous lupus glomerulonephritis. The patient was initially treated with pulse dose steroids and plasmapheresis, followed by cyclophosphamide. He continued steroid therapy post-discharge and was subsequently treated with Rituximab according to the WAVE protocol. His condition improved and he is being followed closely.

Discussion: SLE and ANCA-associated vasculitis, separately, are uncommon rheumatologic diseases. An overlap of these diseases has been reported few times in the literature. We present a unique case of a Caucasian male in his third decade of life, without a previous personal or family history of autoimmune disease, with serological and biopsy findings of both diseases occurring simultaneously. ANCA, typically p-ANCA, can be detected in up to 30% of SLE patients and can be higher with renal involvement. Patients with overlap syndrome have increased complications and higher mortality rates than those with either disease alone.

Our patient was found to have necrotizing and crescentic glomerulonephritis, most consistent with ANCA vasculitis, specifically microscopic polyangitis with MPO positive staining. The biopsy also revealed abundant immune-complex deposits consistent with WHO class V diffuse membranous lupus glomerulonephritis. These diseases are typically seen in young to middle aged females, and given the rarity of this case, biopsy findings were confirmed by two pathologists from separate institutions.

Presentations of autoimmune diseases and vasculitis can be multi-systemic. Immediate action and appropriate work up with a multidisciplinary team is required for optimal patient care. Our patient displayed pulmonary-renal involvement in addition to systemic features such as fevers, myalgia and profound anemia. Considering overlap syndromes, especially in patients with underlying connective tissue disease or systemic vasculitis, is vital for the prevention of excess morbidity in this population.
Hemophagocytic Lymphohistiocytosis Secondary to Babesiosis

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Introduction: Hemophagocytic Lymphohistiocytosis (HLH) is characterized by acute uncontrolled proliferation of lymphocytes and macrophages leading to inappropriate immune activation. HLH may be familial or secondary to infectious, neoplastic, or metabolic causes, often posing a dilemma to identify the underlying etiology.1

Case Presentation: A 38-year-old Caucasian male with type 1 diabetes presented with anuria, acholic stools, jaundice, scleral icterus and arthralgias. He was febrile, had a blanching, petechial rash and diffuse tenderness on the lower extremities. Remarkable labs included hemoglobin 10.2 g/dL, platelets 43,000/mm3, total bilirubin 12.1 mg/dL, and conjugated bilirubin 8.6mg/dL. ALT, AST and alkaline phosphatase were also elevated to 160 IU/L, 192 IU/L, and 286 IU/L, respectively. Haptoglobin was low suggesting hemolysis. Peripheral smear revealed inclusion bodies in a small proportion of neutrophils. Abdominal CT imaging showed mild splenomegaly and hepatomegaly. The patient had a markedly elevated ferritin of 32,383 µg/L and soluble interleukin-2 receptor (sIL2R) of 7,274 U/mL. Coombs testing for anti-C3D direct antiglobulins returned positive. Due to downtrending hemoglobin, as low at 7 g/dL, a bone marrow biopsy was preformed and illustrated hemophagocytosis. At this time six of the eight criteria for the diagnosis of HLH were identified, including fever, splenomegaly, anemia/thrombocytopenia, elevated ferritin, tissue demonstrating hemophagocytosis, and an elevated sIL2R. The patient underwent treatment with the HLH-2004 protocol which included dexamethasone and cyclosporine. Etoposide was held in light of the hyperbilirubinemia. Given the intraerythrocytic inclusions on peripheral smear, PCR testing for parasitic DNA was performed and he tested positive for Babesia microti DNA on Wright staining with 2.8% parasitemia. He was started on azithromycin and atovaquone and his clinical status improved. Upon discharge he was continued on dexamethasone and cyclosporine; he did not require etoposide for resolution of symptoms. Patient completed a cyclosporine and dexamethasone taper and at the 2 month follow up appointment his lab values had normalized.

Discussion: This is the first reported case of a healthy immunocompetent adult less than age 40 old who developed HLH secondary to Babesia. Three previous cases of HLH secondary to Babesia were reported in elderly immunocompetent patients.2 One additional case was reported in 1986 in a 54-year old male who did not have any reported history of immunosuppression.3 In our case, as well as in the previous four reported cases of HLH secondary to babesiosis, significant clinical improvement with symptom resolution occurred within 3-7 days of babesia treatment initiation. It is important to be aware of the diverse presentation of HLH that can allow for the early detection and treatment of the underlying condition which may help improve clinical outcomes.

References
Presumed Lymphocytic Hypophysitis in a nonpregnant patient presenting with Hypopituitarism

Authors: Nirmal Nair, Leigh Kwak

Introduction: Lymphocytic hypophysitis is a rare disorder that presents very similarly to primary pituitary adenomas as well as pituitary failure in pregnant women and for this reason it is crucial to keep this possibility within the differential diagnosis when working up a patient presenting with signs and symptoms of hypopituitarism.

Case Presentation: Patient is a 60-year-old woman with no past medical history who was referred to Endocrinology by her primary care physician for workup of acute onset fatigue. Symptoms began in May 2018 with severe fatigue, anorexia, and orthostatic dizziness. The constellation of symptoms prompted workup for primary adrenal insufficiency. Initial labwork showed decreased thyroid hormone production, gonadotropins, and cortisol levels with a cosyntrropin stimulation test showing minimal response. MRI showed enhancement along the pituitary infundibulum and hypothalamus. These findings suggested acute onset hypothyroidism, 5mg oral prednisone was started. She reported symptomatic improvement, but developed daytime polyuria following initiation of steroids and as a result was started on DDAVP as adrenal insufficiency often masks Diabetes Insipidus.

Oncologic workup was done to rule out the possibility of lymphoma. LP was performed to rule out underlying malignancy. Although negative for malignant cells, this showed elevated lymphocytes. In light of increased lymphocytes on lumbar puncture, symptomatic improvement on steroids, and improvement in pituitary enhancement on repeat imaging, it was concluded that this was a case of lymphocytic hypophysitis. Most recent MRI from October 2018 showed continued decrease in enhancement of pituitary stalk compared to the previous MRI.

Discussion: Lymphocytic hypophysitis is a rare condition where the pituitary gland becomes infiltrated by lymphocytes resulting in pituitary enlargement and impaired function. Patients present clinically with signs and symptoms similar to that of symptomatic pituitary adenomas. If pituitary involvement is significant enough to cause mass effect on the optic chiasm or cavernous sinus, patients may additionally present with diplopia and abnormal pupillary exam1.

Initial evaluation is focused on establishing hormonal deficits localized to either posterior or anterior pituitary, including TSH, growth hormone, and LH and FSH levels in post-menopausal women. Urine osmolarity is obtained in order to rule out Diabetes Insipidus. An MRI with and without contrast shows diffuse enlargement of the pituitary with a homogeneous appearance. Owing to the similarity of this condition both in presentation and in some cases on imaging with pituitary adenomas, the gold standard for diagnosis is biopsy2.

The initial goal of treatment for lymphocytic hypophysitis is focused on reducing inflammation with corticosteroid therapy as well as hormonal replacement. Estimates suggest that more than 70 percent of patients with this condition may require life-long replacement3. Our patient is fortunate in that with physiologic steroids there was some resolution but the need for replacement still exists as when the DDAVP was inadvertently held, the polyuria returned.

References

Fluid overload or could it be something else?

Authors: Saketh Parsi, M.D. Lakshmi Priyanka Mahali, M.D.

Introduction: Diagnosis of RA myxoma should be suspected in patients with symptoms mimicking congestive heart failure associated with weight loss.

Case Presentation: A 71 year-old woman with hypertension presented with progressive ascending bilateral leg swelling for one week. She also reported chronic dry cough, wheeze and dyspnea on exertion since over one month and unintentional weight loss of 40 pounds since over a year. She was hemodynamically stable. Physical exam revealed a diastolic murmur on the left sternal border. Transthoracic Echocardiography (TTE) reported severe right atrial dilatation and a pedunculated, mobile mass with multiple stalks (3.1 x 1.9 cm in longest dimensions) attached to interatrial septum, consistent with an atrial myxoma. Computer tomography (CT) of chest revealed bilateral extensive pulmonary embolisms (PE), with right heart strain, pulmonary arterial hypertension, and irregular filling defect within the right atrium (RA) corresponding with the myxoma. Anti-coagulation (AC) was started with lovenox and patient was discharged on apixaban with cardiothoracic clinic follow up.

Discussion: Approximately 80 percent of myxomas are found in the left atrium; the remainders are in the RA. RA myxomas grow into the lumen and obstruct flow, producing hemodynamic changes that are similar to tricuspid stenosis. RA myxomas typically present with signs and symptoms of right heart failure (peripheral edema, hepatomegaly, and ascites) and cause diastolic murmurs. Constitutional symptoms like malaise, anorexia, fever, arthralgia, weight loss are more common in cases of cardiac myxomas due to the release of cytokine interleukin-6 (IL-6).

In addition, tumor fragments may be released into the pulmonary circulation causing symptoms consistent with pulmonary embolism, as seen in our patient. Presence of an atrial septal defect results in shunting of venous blood into the systemic circulation causing hypoxemia or systemic emboli.

Provoked PE is temporarily managed with AC and permanently managed by treating the cause. RA myxomas require prompt resection to prevent further embolization or cardiovascular complications, including sudden death.
Lactobacillus jensenii infection: a myth of being a friendly bacteria

Authors: Ranchal, Purva; Gupta, Rahul; Lobo, Stephen; Goldberg, Randy; El Khoury, Marc

Introduction: Lactobacillus species (sp) constitute a significant component of the microbiota in different parts of the human body including digestive, urinary and genital system. Invasive infections secondary to Lactobacillus sp. have been occasionally reported in immunocompromised individuals. We describe a case of persistent Lactobacillus bacteremia and possible endocarditis associated with a large prostatic abscess in a diabetic patient.

Case Presentation: A 57 year old man with history of hypertension presented with 4 weeks history of progressive malaise, subjective fever and night sweats associated with nocturia without symptoms of dysuria or hematuria. Physical exam was significant for tachycardia 140 beats/minute and temperature 102.5oF. Laboratory findings were pertinent for a WBC of 12.7 k/mm3, blood glucose 652 mg/dL, anion gap 21, HgA1c > 14. Urinalysis showed significant pyuria and bacteriuria. Chest X-ray was unremarkable. He was started on intravenous hydration, insulin drip, cefepime and vancomycin. Urine culture was reported as normal urogenital flora, later speciated as Lactobacillus sp. Blood cultures grew Lactobacillus sp in one out of 4 bottles which was initially considered a contaminant. A week later, patient remained febrile and subsequent two sets of blood cultures also grew Lactobacillus sp. A contrast CT scan of the chest, abdomen and pelvis showed multiple large abscesses within the seminal vesicles and prostate extending into the right obturator internus muscle with incidental findings of bilateral peripheral cavitary lung nodules consistent with septic emboli concerning for right side endocarditis. Antibiotics were switched to ampicillin. CT guided drainage of the prostatic abscess was performed and abscess cultures grew Lactobacillus sp. identified as Lactobacillus jensenii using a 16S-RNA universal PCR. Patient received 8 weeks of IV ampicillin followed by oral amoxicillin until repeat CT scan of pelvis showed interval resolution of the prostate abscess 3 months later. A transesophageal echocardiogram showed no valvular vegetation.

Discussion: Lactobacillus sp. an important commensal of the human microbiome, are considered a human friendly bacteria, rarely causing infections. Prostate abscess caused by L. jensenii has not been reported before. There are few case reports on bacterial infections such as endocarditis caused by L. jensenii. This case describes the extent of sepsis caused by Lactobacillus. The presence of septic pulmonary emboli in this case are either from prolonged significant bacteremia or right sided endocarditis despite negative echocardiogram. Patient responded well to drainage of the abscess and prolonged course of ampicillin. Since the morbidity and mortality associated with Lactobacillus infections is high, it is important to differentiate true bacteremia from contamination and use of appropriate antimicrobials.
Granulomatosis with polyangiitis presenting as a large bleeding pleural based mass

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Introduction: Granulomatosis with polyangiitis (GPA), is a systemic necrotizing vasculitis, which affects small and medium sized blood vessels and is often associated with cytoplasmic anti-neutrophilic cytoplasmic antibodies (c-ANCA). Characteristically, it presents with the triad of upper respiratory tract, lower respiratory tract and kidney involvement. However, it can present with lung masses or nodules which can be mistaken for neoplastic processes.

Case Presentation: A 69-year-old man presented with abdominal pain and nausea of one day’s duration. CT abdomen did not show any abdominal pathology but revealed incidental multiple lung nodules. CT Thorax also showed multiple small nodules bilaterally with a dominant 3.9 x 1.7 cm pleural based mass in the right lower lobe (RLL). Initial suspicion was for metastatic malignancy and the patient underwent an endobronchial ultrasound (EBUS) with biopsy of the RLL mass that showed small fragments of benign bronchial wall with non-specific fibrosis and mild eosinophilic infiltrate. His symptoms resolved with conservative management and he was discharged home. One week later, he presented with severe hemoptysis that lead to hemorrhagic shock. CT Angiogram of the thorax showed increase in the size of the lesions seen on previous imaging. Repeat EBUS revealed a large clot in his RLL mass with blood slowly oozing around it and he underwent IR-guided embolization of a small pseudoaneurysm in that mass. On further review it was found that the patient had a history of recurrent sinus infections so an autoimmune workup was initiated for concern of vasculitis. The patient was found to have a positive c-ANCA titer of 1:360, with a markedly elevated anti proteinase 3 antibody titers and a negative anti MPO antibodies. ESR was elevated at 66 and CRP was elevated at 361. He was diagnosed with ANCA-associated vasculitis, specifically GPA and started on high dose steroids with subsequent improvement of his symptoms. After discharge he received Rituxan with further improvement in his lung nodules.

Discussion: GPA can mimic multiple lung diseases on chest imaging including pneumonia, emboli or neoplastic processes. It is not uncommon to have lung masses or nodules which can be asymptomatic or can present with life-threatening bleeding similar to our patient. Evaluation for GPA should be considered in patients with respiratory symptoms and lung masses in the absence of malignancy and infectious etiology.
Extremely Elevated Eos: A Case of Levetiracetam-Induced Eosinophilia

Authors: Masha J. Slavin, MD and Todd S. Cutler, MD

Introduction: Cases of severe eosinophilia require investigation into the etiology of the eosinophilia. In the hospitalized patient, medication-induced eosinophilia should be considered. Work-up for end-organ damage is crucial to assess for both clinical and subclinical effects. We present the case of a woman with levetiracetam-induced eosinophilia.

Case Presentation: A 67-year-old woman with breast cancer (s/p mastectomy, on chemotherapy with no evidence of disease) and deep vein thrombosis on rivaroxaban presented to an outside hospital with 1 week of worsening confusion. She was started on levetiracetam for seizure activity on electroencephalogram, and the dose increased later in the hospitalization. Her confusion persisted, and she was transferred to our institution for additional work-up, which yielded a diagnosis of autoimmune encephalitis that improved after 5 plasma exchange treatments. While hospitalized, she completed a course of piperacillin-tazobactam and cefazolin for E. coli bacteremia from a urinary source and ampicillin-sulbactam for aspiration pneumonia.

Patient was clinically asymptomatic awaiting discharge, not requiring daily blood draws. When labs were again checked, she was noted to have eosinophilia (48.3%, absolute eosinophil count 3,600/uL), with normal WBC, and hemoglobin and platelets near her baseline. Eosinophils peaked at 74%, absolute count 10,500/uL, even causing a neutropenia. On further review, her eosinophils were up trending at the outside hospital, but had normalized after she received methylprednisolone for the autoimmune encephalitis.

Patient remained asymptomatic and work-up for end-organ damage was negative, including a normal chest x-ray, urinalysis, creatinine, troponin, and liver chemistries. She had normal morning cortisol and negative Strongyloides IgG. There was low suspicion for development of a new hematologic disorder or malignancy during her hospitalization. Her antibiotics had been completed before the eosinophils again started to rise. The etiology was thought to be due to levetiracetam, which was rapidly down-titrated and discontinued, with improvement in her eosinophilia in the coming weeks.

Discussion: Etiologies of eosinophilia include malignancy and hematologic disorders, parasites such as Strongyloides, and hypoadrenalism. Several medications are known to cause eosinophilia. There have been case reports of levetiracetam causing eosinophilic pneumonia, as well as drug rash, eosinophilia, and systemic symptoms (DRESS). Cephalosporins are also associated with eosinophilia, including eosinophilic pneumonia. Because the main concern with eosinophilia is the possibility of end-organ damage, patients should be assessed for a broad range of symptoms. For instance, eosinophilic involvement of the skin can cause a rash, involvement of the respiratory system may lead to cough, and cardiac involvement may cause signs of myocarditis. Because end-organ effects of eosinophilia can be subclinical, work-up should include a chest x-ray, urinalysis, creatinine, liver chemistries, and troponin. Treatment for eosinophilia involves addressing the underlying cause (e.g., medication discontinuation, treatment of parasite infection). With severe eosinophilia, steroids may be helpful. However, even with treatment, eosinophilia can take months to resolve.

References

NEW YORK CLINICAL VIGNETTE POSTER FINALIST - BASEER SYED, MD

A Rare Case of Sarcoidosis: Anemia and Acute Renal Failure

Authors: Baseer Syed MD, Ajay Tambe MBBS, Kunal Bhuta MD, William DiFilippo MD

Introduction: Sarcoidosis is an inflammatory disorder characterized by the presence of non-caseating granulomas in affected organs. It primarily involves the lungs though it may also involve other organs such as the skin, eyes, liver, bone marrow, spleen and kidney. Renal impairment is seen in 4-20% of cases though granulomatous interstitial nephritis (GIN) is only observed in less than 1%. Bone marrow involvement is reported in less than 5% of cases. It may present as anemia, leukopenia, thrombocytopenia or deficiency of all three-cell lines due to granulomatous infiltration. Renal and bone marrow involvement, without pulmonary features, is extremely rare and can make the diagnosis challenging.

Case Presentation: A 27-year-old African-American female presented with complaints of dyspnea, malaise and 40 pound weight loss over past few months. Her history was significant for gastroesophageal reflux disease (GERD), Grave’s disease, and an upper respiratory tract infection 6 months prior to presentation that was attributed to streptococcal pharyngitis. Her medications included Levothyroxine and Protonix. On physical examination she was found to be tachycardic. Lab studies revealed a hemoglobin 7.2 g/dL and serum creatinine 4.4 md/dL. Thyroid stimulating hormone (TSH) and calcium were normal. Urinalysis had a low specific gravity only and urine sediment was bland. A full hemolysis work up was negative. Stools were guaiac negative. A comprehensive Vasculitis work up was also negative. Renal ultrasound and CT scan of the abdomen showed large kidneys and no other structural defects. Chest radiograph and CT of chest were normal. Infectious workup was significant for positive parvovirus IgG. HIV was negative. Serum and urine electrophoreses were negative as well. Serum calcium was normal.

She was treated conservatively with hydration and discontinuation of her protonix. Her anemia progressed to a Hb of 6.7 mg/dL. The Serum creatinine stabilized at 3.5 mg/dL. EGD was normal. A bone marrow biopsy was performed and revealed sheets of non-caseating granulomata. An ACE level was normal. A renal biopsy followed and revealed granulomatous interstitial nephritis (GIN) and fibrosis with mild tubular atrophy. The patient was given oral prednisone (60 mg/day) with significant improvement in her renal dysfunction and anemia two weeks following the start of therapy.

Discussion: Renal involvement in sarcoidosis is most often due to nephrocalcinosis, hypercalciuria or calculi secondary to hypercalcemia caused from increased vitamin D production by activated macrophages in the granulomata. GIN is usually silent and rarely causes significant renal impairment. Serum ACE is not reliable in the diagnosis with a reported 60% sensitivity. This case highlights an atypical and extremely rare presentation of non-pulmonary sarcoidosis. Systemic corticosteroids continue to be the mainstay of treatment in this unpredictable disease.
NEW YORK CLINICAL VIGNETTE POSTER FINALIST - BRYAN E-XIN TAN, MD

Bilateral Adrenal Hemorrhage: An Underrecognized Complication Of Heparin-Induced Thrombocytopenia

Authors: Bryan E-Xin Tan, Muhammad Waqas Tahir, Qamar Ahmad, Department of Internal Medicine, Rochester General Hospital, Rochester, NY

Introduction: Heparin-induced thrombocytopenia (HIT) is a well-known complication of treatment with heparin. Thrombocytopenia is the most common manifestation of HIT, but it can cause catastrophic arterial and venous thrombosis with a mortality as high as 20 percent. Bleeding is rarely seen in HIT but has been reported. We present a case of HIT with portal and splenic vein thrombosis, found to have bilateral adrenal hemorrhage.

Case Presentation: A 63-year-old female presented with abdominal pain, nausea, vomiting, and constipation. CT scan showed evidence of sigmoid volvulus, and she underwent subtotal colectomy and was subsequently discharged without any complications. She was readmitted due to persistent abdominal pain and X-ray was consistent with paralytic ileus. The patient was managed with supportive treatment. She spiked a fever on day six. Cefepime was started for possible intra-abdominal infection. On day nine, CT scan of the abdomen revealed splenic and portal vein thromboses, and bilateral adrenal gland enlargement consistent with bilateral adrenal hemorrhage. She spiked a fever again and became hypotensive. Lab work revealed a drop in platelet count to 122,000 from 328,000 at time of admission. Her 4T score was calculated to be 7 indicating a high probability of HIT and her morning cortisol level was 1.8µg/dL consistent with adrenal insufficiency. She was started on IV dexamethasone, and all heparin products were discontinued. HIT ELISA assay was positive for anti-platelet factor 4 (PF4)-heparin antibody, and a diagnosis of HIT was made. She was started on intravenous bivalirudin and was later anticoagulated with warfarin. She had an uneventful hospital course later on and was discharged home with warfarin, fludrocortisone, and hydrocortisone.

Discussion: HIT causes anti-PF4-heparin antibody mediated thrombocytopenia resulting in acquired hypercoagulability syndrome which leads to arterial and venous thrombosis. Hemorrhage is uncommon in HIT but has been reported. In a series of 6332 patients hospitalized with HIT, bleeding was seen in 6 percent of patients. This study did not report whether bleeding was due to thrombocytopenia from HIT or to anticoagulation for HIT treatment.

Interestingly, our patient was found to have both venous thrombosis and bilateral adrenal hemorrhage. Bilateral adrenal hemorrhage in HIT could be due to thrombocytopenia itself or as a result of acquired hypercoagulability syndrome. The distinctive vascular architecture makes adrenals vulnerable to adrenal vein thrombosis complicated by hemorrhagic transformation. Ironically, the hemorrhage is treated as a thrombotic disorder, and patients require anticoagulation and adrenal replacement therapy.

The complication of adrenal vein thrombosis leading to bilateral adrenal hemorrhage remains underrecognized and undertreated. Unrecognized adrenal hemorrhage secondary to HIT can lead to significant mortality due to adrenal collapse. Clinicians should maintain a high index of suspicion to achieve prompt diagnosis and provide life-saving intervention.
IMMUNOSUPPRESSION, LUPUS, AND PRIMARY CNS LYMPHOMA

Authors: Laxmi Upadhyay MD; Alzira Avelino MS-IV; Prasanta Basak MD; Kameshowri Lakshmi, MD; Stephen Jesmajian, MD, Department of Medicine, Montefiore New Rochelle Hospital and Albert Einstein College of Medicine, New Rochelle, New York.

Introduction: The use of immunosuppressive therapy, particularly mycophenolate mofetil (MMF) and azathioprine has been associated with the development of primary central nervous system lymphoma (PCNSL). PCNSL is well studied in patients with acquired immune deficiency syndrome or organ transplantation. However, it is rarely diagnosed in rheumatic diseases. Scattered cases of immunodeficiency related PCNSL have been reported in patients with rheumatoid arthritis, myasthenia gravis, and Crohn’s disease. We present a case PCNSL in a SLE patient treated with MMF.

Case Presentation: A 50-year-old African American female with SLE presented with intermittent left temporal pressure like headache for 2 weeks. She also had decreased appetite, fever and fatigue. Earlier she had been diagnosed with a breast tumor, hypertension, hyperlipidemia and chronic kidney disease secondary to lupus nephritis. She was on low dose prednisone for 10 years and MMF 1000mg twice a day for the last 5 years. On examination she was alert and oriented, T 39.3 degree Celsius, BP 151/69 mmHg, heart rate 82/min, respiratory rate 16/min, and oxygen saturation 100% on room air. Physical exam was unremarkable. WBC 6.5 with 13% monocytes, C3 and dsDNA were normal. CT head showed new low attenuation abnormalities involving the left parietal periventricular white matter with new mass effect on the left lateral ventricle. She was started on dexamethasone, prednisone and MMT discontinued. MRI of the brain with contrast showed ring-enhancing lesion involving the left periventricular white matter along with dura matter involvement along the right temporal lobe. CT scan of the chest/abdomen/pelvis was normal. Tumor markers including CA 27.29, CEA, CA-125, CA 19-9 were negative. Tests for HIV, TB, syphilis and Cryptococcus were also negative. Lumbar puncture revealed normal opening pressure, 5-8 WBCs, normal glucose and protein, negative for enterovirus RNA, cryptococcal antigen and toxoplasma antibody. CSF PCR was positive for cytomegalovirus and EBV. Brain biopsy showed patchy subcapsular and perivascular lymphoid infiltration composed of intermediate to large sized cells with irregular nuclei contour, vesicular chromatin, distinct nucleoli and moderate amount of eosinophilic cytoplasm. Immunoperoxidase studies demonstrated atypical cells positive for CD20 compatible with EBV-associated B cell lymphoproliferative disorder, and negative for CMV, CD3, CD5, BCL6. The patient was discharged on dexamethasone.

Discussion: Initial differentials for the CT findings included infection, lupus cerebritis and metastasis from breast cancer. The brain biopsy was diagnostic for PCNSL. There has been a growing association between lymphoproliferative disorders, PCNSL and the use of MMF. These patients may benefit from MMF withdrawal and treatment with rituximab. Until recently, only 8 cases of SLE accompanied with PCNSL were reported. We highlight the possibility of PCNSL with immunosuppression in SLE and emphasize the importance of expedited imaging with CNS symptoms even without any focal neurological signs.
What a Fluke! The first documented case of cholecystic varices causing biliary obstruction as a sequelae of schistosomiasis

Authors: Yip, Kevin; Cheng, Austin; Swanenberg, Irene, Department of Internal Medicine, New York University School of Medicine, New York City

Introduction: We present the first documented case of cholecystic varices causing extrinsic biliary obstruction due to schistosomiasis sequelae. Although rare, it is a possible cause of hyperbilirubinemia which can be treated successfully with stent placement. Chronic schistosomiasis infection should be considered for patients from endemic regions presenting with portal hypertension and cirrhosis.

Case Presentation: A 53-year-old Sierra Leonean man with schistosomiasis presented with two months of jaundice and abdominal distension. Two years earlier, he was diagnosed with schistosomiasis complicated by non-cirrhotic portal hypertension, splenomegaly, and esophageal and gastric varices. Initially on this presentation, this patient was septic due to MDRO E.Coli bacteremia secondary to spontaneous bacterial peritonitis and was newly diagnosed with cirrhosis attributed to chronic schistosomiasis.

Notably, this patient presented with common bile duct obstruction with total bilirubin of 27mg/dL (direct bilirubin 12.4mg/dL) caused by extrinsic compression by a large cholecystic varix encircling the narrow portion of the common bile duct. These ectopic varices are a distinctly uncommon complication of portal hypertension, confirmed by both MRCP and ERCP. The patient’s hyperbilirubinemia resolved within two days after common bile duct stent placement.

Discussion: Obstructive jaundice is a common clinical scenario, however extrinsic compression of the common bile duct by a cholecystic varix is a rare entity. This patient’s presentation of post-hepatic obstructive jaundice with severe hyperbilirubinemia was similar to that of more prosaic processes such as common bile duct intrinsic or extrinsic obstruction by stone or pancreatic head mass, for instance. Gallbladder varices form due to collaterals between the cholecystic and pericholecystic venous systems which are usually associated with portal vein thrombosis. Similar to esophageal or gastric varices, the pathophysiology of cholecystic varices involves portal hypertension leading to dilation and reversal of blood flow in these portal vessels. Our case suggests that it may be prudent to perform MRCP prior to ERCP or other interventions to evaluate for atypical varices.

Additionally, our case highlights chronic schistosomiasis, a known but rare cause of cirrhosis. If untreated, the migration and subsequent entrapment of eggs in various organ systems leads to the late phase of disease, often years after initial exposure. Hepatosplenic involvement is common in chronic infection from any of the main Schistosoma species, specifically following entrapment of schistosome eggs in the portal vessels causing inflammation and collagen deposition. This fibrosis and subsequent portal hypertension are distinct from most etiologies of cirrhosis, which are primarily hepatocellular processes. Early diagnosis and treatment of schistosomiasis is therefore imperative to avoid these late sequelae.

References

Stroke-like Presentation in a Young Adult: PML

Authors: Aiman Zafar, Mukesh Kumar and Marilou Corpuz. Department of Medicine Montefiore Medical Center, Bronx, New York

Introduction: Strokes in patients less than 40 years old are uncommon except for patients with underlying risk factors for atherosclerotic heart disease. Other causes include vasculitis or infections such as syphilis. We present a patient with clinical and radiological manifestations of a stroke who was found to have progressive multifocal leukoencephalopathy (PML).

Case Presentation: A 32 YO man with no prior medical history presented with numbness, tingling and weakness in his left arm, slurred speech and facial muscle weakness for 1 week. His exam showed left sided facial droop, weakness of left upper extremity with decreased sensations and hyperreflexia. MRI brain showed sub-acute to chronic infarcts involving right frontal lobe, centrum semiovale, corona radiata and posterior watershed territory, hence, acute stroke was diagnosed. Extensive work up was done. He was positive for HIV 1 with a CD4 count of 73 cells/uL. Lumbar puncture was positive for JC polyoma virus PCR (3588 copies/ml). Patient was suspected to have PML although his neuro imaging features were not classic PML findings. Anti-retroviral therapy (ART) was initiated. Few weeks later he was readmitted with seizures. A contrast MRI brain performed few months later revealed findings that were more consistent with PML. A follow up MRI remained stable and the patient showed clinical improvement in his symptoms.

Discussion: PML is a severe, progressive and fatal demyelinating disease of the central nervous system caused by reactivation of the JC polyomavirus in immunosuppressed individuals. It is now increasingly recognized in patients with HIV and is an AIDS defining illness. It is seen in advanced stages of HIV disease with CD4 count <200 cells/uL and can also herald the manifestation of HIV. Clinical features overlap with stroke and include altered mental status, hemiparesis/monoparesis, limb/gait ataxia, hemianopia and diplopia. Seizures are often seen. On neuroimaging, the lesions are bilateral, asymmetric, multifocal, preferentially involving the periventricular areas and subcortical white matter of frontal/occipital/parietal lobes and do not exhibit mass effect or contrast enhancement. On CT PML lesions appear as hypodense patchy or confluent white matter regions and on MRI brain as areas of increased signal on T2-weighted images and fluid attenuated inversion recovery (FLAIR) sequences. Although brain biopsy is gold standard for making a diagnosis, the presence of JC virus DNA in CSF using PCR and clinical and neuroimaging findings are sufficient to diagnose PML. There is no specific therapy other than improving host’s immune response via continuation or initiation of an effective ART.

Since PML symptoms can overlap with stroke and initial neuro imaging findings can also be misleading, it is crucial to consider HIV testing in a young patient with no known risk factors presenting with stroke like symptoms. High clinical suspicion in the right clinical context is imperative to proper diagnosis.
Right Ventricular Hypertrophy and Right Heart Failure Secondary to Partial Anomalous Pulmonary Venous Connection

Authors: Sumaira Zareef MD, MPH, Sangeeta Venughopal MD, Keron Lezama MD, Montefiore Medical Centre, Department of Internal Medicine.

Introduction: Anomalous pulmonary venous connection is an uncommon congenital malformation that can be categorized into partial or total, of which the former is more common. Partial anomalous pulmonary venous connection (PAPVC) has been implicated as a cause of right ventricular hypertrophy (RVH) and right heart failure (RHF) however is often overlooked in the diagnostic work up of RVH and RHF.

Case Presentation: Our patient was a 71-year-old nonsmoking woman with history of mitral valve prolapse, and hypertension who was admitted for worsening dyspnea, fatigue, decrease in exercise tolerance (ET) from unlimited at baseline ET to one block, and melena for two weeks. The working diagnosis was anemia as her hemoglobin on admission was 13 g/dl but extent of her symptoms seemed out of proportion to her mild anemia. While undergoing work up of anemia and melena with esophagoduodenoscopy (EGD), no abnormality was detected but she became hypoxic with oxygen saturation of 85% during the procedure. Ventilation perfusion scanning did not reveal any VQ mismatch. Transthoracic echo (TTE) revealed moderate right atrial dilatation, right ventricular hypertrophy, moderate to severe tricuspid regurgitation (TR), and no atrial septal defect (ASD). These findings were also present on TTE done one year before the current hospitalization. CT chest was done to explore further the etiology of hypoxic event at end of EGD. CT chest did not identify any pulmonary pathology but revealed PAPVC and was further confirmed by congenital heart disease echo. At her most recent cardiology visit after discharge she had NYHA class 2 heart failure, could do most of activities of daily living and refused any catheter based surgical repair.

Discussion: Many patients with PAPVC remain asymptomatic, however some may present at a later age with symptoms related to pulmonary hypertension and RHF. PAPVC initially acts as a left to right shunt causing the oxygenated blood to be returned to right heart without entering the systemic circulation and does not cause any symptoms. Overtime, increasing pulmonary blood flow causes increased pulmonary vascular resistance, pulmonary hypertension, and right ventricular overload leading to right heart remodeling and enlargement. Right ventricular hypertrophy and pulmonary HTN can cause severe TR and right atrial arrhythmias which can cause sudden clinical decompensation. RHF can develop over time. If an atrial septal defect is present, shunt reversal (now predominantly right to left) causes systemic cyanosis and Eisenmenger syndrome. Our patient did have right ventricular dilatation on TTE done one year ago but further diagnostic work up was not done most likely due to asymptomatic nature of her anomaly. Physicians should keep PAPVC on their list of differentials as a cause of dyspnea with right ventricular dilatation in order to intervene early and to avoid any clinical complications.
Intussusception as the first manifestation of HIV-AIDS

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Introduction: Intestinal intussusception is defined as the telescoping of a segment of the gastrointestinal tract within the lumen of the adjacent segment and presents with mechanical bowel obstruction (BO). Intussusception is rare in adults, accounting for only 1 to 5 percent of BO. Here we describe a young man who presented with intussusception secondary to a primary B-cell Lymphoma of the Ileum on the background of a yet undiagnosed underlying Acquired Immunodeficiency Syndrome (AIDS).

Case Presentation: A 31 year old Nigerian man with no known medical history was admitted to our hospital with persistent nausea, vomiting and crampy abdominal pain of 5 days duration. He also complained of abdominal distention with constipation. On physical examination, he was afebrile and had mild oral thrush. Abdomen was distended and tender to palpation and breath sounds were decreased on the right side. Computed tomographic scan of chest, abdomen and pelvis with contrast showed small bowel obstruction secondary to an ileocolic intussusception, a 2.4cmx2.3cm hypodense mass in the left lobe of the liver and an 8cmx5.6cm lung mass in the right middle lobe. Patient underwent urgent exploratory laparotomy with right hemicolectomy including the terminal Ileum and biopsy of the liver mass. Post-operative period was complicated by low grade fevers unresponsive to antibiotics. Urine and blood cultures, RPR and TB-quantiferon were tested negative. His labs showed persistent neutropenia prompting HIV testing which came back positive for HIV-1 infection. His CD4 count was 87cells/mcl. Histology of the terminal ileum and liver mass biopsy was consistent with Diffuse Large B-Cell Lymphoma. Patient was started on HAART and Bactrim prophylaxis. Oncologist recommended chemotherapy as outpatient. He was subsequently discharged.

Discussion: Intussusception is a common cause of BO in children as compared to adults. Intussusception develops typically due to a pathologic lead point within the bowel, secondary to the presence of intra- or extra-luminal lesions (inflammatory lesions, Meckel's diverticulum, postoperative adhesions, lipoma, polyps, lymphoma and metastases). The lead point is pulled forward by normal peristalsis, telescoping or prolapsing the affected segment of bowel (intussusceptum) into another segment of bowel (intussuscipiens).

An increased incidence of intussusception has been reported in AIDS patients and is likely attributable to the high incidence of infectious and neoplastic conditions of the bowel in these patients, such as lymphoid hyperplasia, Kaposi's sarcoma, non-Hodgkin's lymphoma and cytomegalovirus (CMV) colitis. AIDS patients presenting with intussusception have been described in the literature, but only two cases have been described where intussusception secondary to a gastrointestinal AIDS defining malignancy lead to the diagnosis of AIDS. Our patient’s neutropenia and oral thrush pointed towards an immunosuppressive state and we emphasize the importance of considering HIV testing in eligible young patients with intussusception.
Point of Care Ultrasound Defuses a Ticking Time Bomb: Early Bedside Evaluation of Infective Endocarditis

Authors: Felix Nau Jr. M.D. Ph.D., Asish Gulati D.O., Christopher Kelly M.D., Jamie Kowal D.O., Chaudry Majeed M.B.B.S.

Introduction: Point of Care Ultrasound (POCUS) is becoming an invaluable tool in the evaluation of the acutely ill patient. However, the ability to assess for certain conditions with POCUS is directly related to the extent of each individual’s training. As such, formal POCUS training by means of a "Teach the Teacher" model has been implemented at Wake Forest Baptist Medical Center (WFBMC), which has led to increased integration of this technology in routine patient care. This case exemplifies how cardiac POCUS expedited a preliminary diagnosis and directly influenced early initiation of appropriate treatment.

Case Presentation: A 30 year-old female presented one day following a normal vaginal delivery from an outside hospital after chest CT angiography (CTA) identified a 2.5 cm pericardial effusion. Review of systems was positive for dry cough, left sided chest pain, dyspnea on exertion, recent post-partum abdominal pain, chills, and diaphoresis. On arrival she was afebrile, BP 140/83 without pulsus paradoxus, HR 100 bpm, RR 20, and normal O2 saturation. Physical exam revealed rales at the lung bases, bilateral pedal edema, and JVD to angle of the mandible. Labs were significant for a WBC 22,700, procalcitonin of 1.78, ESR 52, CRP 128, BNP of 587, and troponin peak of 0.110. EKG showed no ST changes and was without electrical alternans. On initial evaluation, a bedside POCUS exam revealed a vegetation approximately 2 cm in diameter on the mitral valve concerning for infective endocarditis (IE). As a result, cultures were performed and the patient was immediately started on empiric broad spectrum antibiotics. TEE further confirmed the presence of a large vegetation with multiple echolucencies suggestive of valvular abscess. Blood cultures came back positive for Streptococcus mitis/oralis. Cardiothoracic Surgery was consulted and mitral valve repair was performed. The patient was discharged home with 6 weeks of IV ceftriaxone and did well in the outpatient setting.

Discussion: The utility of POCUS in evaluating cardiovascular disease has been examined at length in the inpatient setting. POCUS exams can help manage conditions such as shock, cardiac arrest, acute heart failure and valvular pathology. According to the American Heart Association guidelines, a TEE is the recommended initial test in high-risk individuals with suspected IE. However, a bedside POCUS exam can be an invaluable tool when a formal TTE or TEE is not readily available. We argue here that all Internal Medicine residents should be trained in bedside ultrasound and POCUS should be considered as an extension of the physical exam to further evaluate certain medical scenarios in the acute inpatient setting.

References

Rare Case of Primary Retroperitoneal Dedifferentiated Liposarcoma

Authors: Dr. Fuad Bashjawish, PGY-2, Dr. Maryselle Winters, PGY-2, Dr. Dezarae Leto, PGY-2, Student Dr. Julian Jacob, OMS-III

Introduction: Retroperitoneal sarcomas are extremely rare tumors only affecting 2 to 5 per million population accounting for 0.1% of all malignancies. Liposarcoma is the most common of all retroperitoneal sarcomas responsible for approximately 20% of all sarcomas in adults. Most important prognostic factors are tumor grade, the presence of positive margins, tumor integrity and degree of resection.

Case Presentation: Here we present a case of a rare retroperitoneal sarcoma. Patient is a 73 yo male who presented to our institution with a large abdominal mass. On work up, a CT abdomen/pelvis demonstrated a 15x15cm heterogeneous, left sided intra abdominal mass extending from the inguinal region superiorly to the level of the kidney. Patient underwent radical resection of retroperitoneal tumor, left colectomy and left nephrectomy. Final pathology demonstrated a high grade, dedifferentiated liposarcoma with rhabdosarcomatous component. Post-operative course complicated with a small intraabdominal abscess formation diagnosed on imaging as well as residual tumor in the left abdomen. The abscess was treated with IV antibiotics and patient was discharged home. Patient was then re-admitted for fascial dehiscence, therefore, patient was taken back to the operating room for additional resection of retroperitoneal mass and Hartmann's procedure. Patient was discharged home with follow up with medical and radiation oncology for adjuvant therapy.

Discussion: Achieving a complete resection and the grade of the tumor are the most important prognostic factors for patient survival. It is important to approach this disease process with a multidisciplinary team that includes surgical, medical and radiation oncology. It is important to recognize that the survival rates are inversely proportional to the grade of the tumor. Even with radical resection of the tumor, there is always a risk of residual tumor cells within the tumor bed that contributes to recurrence and additional surgical interventions. Further research into different treatment methods with systemic chemotherapy or novel targeted therapeutic trials may improve the outcomes of these patients.

References

A Rash that Follows None of the Rules: A Unique case of Disseminated HSV-2

**Authors:** Dr. Thomas Blair, MD; Dr. Darryl Chang, MD; Dr. Sowmya Nagaraj, MD

**Introduction:** Herpes simplex virus 2 (HSV-2) infection often presents in a disseminated pattern in immunocompromised hosts. Immunocompetent hosts typically present with genital or oral lesions or have HSV-2 or varicella-zoster virus (VZV) that follows along a dermatomal pattern. Here we present a herpetiform rash, which was culture positive for HSV-2 on biopsy, that disseminated across an immunocompetent patient’s lower extremities after a T1-T6 spinal cord injury. She had no previous known history of HSV-2 and was exposed to VZV as a child with chicken pox, but presented with a new herpetiform rash disseminated across lower extremities and abdomen after neurosurgical repair.

**Case Presentation:** A 40 year old caucasian female was transferred from the surgical ICU to the general internal medicine service for chronic management of medical conditions. The patient had been involved in a motor vehicle accident which involved T1-T6 fracture and had needed chronic tracheostomy management. She had been treated for ventilator associated pneumonia with 2 weeks of Vancomycin and piperacillin/tazobactam and had responded well. However, during hospitalization, a new herpetiform rash was noted with vesicles and erythema across bilateral lower extremities and abdomen across multiple dermatomes without any distinguishable pattern except that it was below the level of her previous injury. She was afebrile, not taking any immune-modulating or immunosuppressive agents, and was not systemically ill. HIV antibody screen was negative. She did not note any symptoms secondary to her lower extremity paraplegia. Antiviral therapy was initiated using acyclovir and a skin biopsy with culture, VZV, HSV1&2 fluorescent antibodies, and HSV cultures were obtained. The patient was scheduled for punch biopsy with a referral to dermatopathologist after antibodies for VZV and HSV were negative, however HSV culture grew HSV-2. Patient then responded to acyclovir and vesicles begin to scab over and erythema decreased significantly.

**Discussion:** This case presents a unique diagnostic challenge of a rash that visually appears consistent with a herpetic form rash that would be consistent with HSV-1 or HSV-2. However, the distribution was not at all consistent with typical HSV-1 presentations and exceedingly atypical for HSV-2. The difficulty in obtaining a diagnosis was compounded by the fact that the system no longer retains inpatient hospital consultation services for Dermatology. While HSV-1 and 2 were of low suspicion given the patient’s immunocompetent status and rash distribution, direct antibody testing and culture were obtained as initial workup. The uniqueness of an immunocompetent patient developing bilateral herpetic rash only and exclusively underneath the level of their spinal injury as the first presentation of HSV-2 without any documented genital lesions has yet to be reported elsewhere. The mechanism of traumatic spinal cord injuries in modulating the immune system’s response in HSV-2 infections is unknown, but this case suggests some interplay.

**References**

Paradoxical bradycardia after nitroprusside infusion: a reminder of the mysterious physiology of the heart

Authors: John Deppe MD, Teresa Campanile MD, Leigh Vidrine PharmD, BCPS

Introduction: Paradoxical bradycardia after nitrovasodilator infusion, although listed in some reference texts, is a clinically underappreciated, physiologically mysterious phenomenon. The Bezold-Jarisch reflex (BJR) provides a rational explanation for the paradoxical bradycardia, rather than tachycardia, in the setting of peripheral vasodilation and relative hypotension. The poorly understood BJR is suspected to represent a cardioprotective reflex during episodes of rapid left ventricular (LV) emptying or LV myocardial irritation. We present the case of a 95-year-old male admitted for hypertensive urgency who experienced sinus bradycardia to 26 bpm and syncope while receiving sodium nitroprusside (SNP) infusion. We believe his case is consistent with a BJR mediated adverse event. The patient recovered fully with cessation of SNP. He had no evidence of an ischemic event and subsequently tolerated nitroglycerin infusion.

Case Presentation: 95-year-old male with prior myocardial infarction, diastolic heart failure, and hypertension was admitted for hypertensive urgency with no end-organ damage. Initial BP 195/41 HR 61. Within 24hrs of admission, he showed poor response to oral antihypertensives and IV labetalol. Acute onset of a headache in setting of sBP 242 with HR 70 prompted his transfer to ICU following a CT head, which was negative for acute hemorrhage. SNP at 0.5 mcg/kg/min was initiated. Nearly instantaneously, his heart rate dropped to 26 bpm with sinus rhythm. He became briefly unresponsive, diaphoretic, and pale. Blood pressure remained 140-180s/50-70s. Discontinuation of SNP normalized his heart rate and mental status within one minute. Subsequent chest x-ray was unremarkable. EKG showed normal sinus rhythm, no ischemic changes, HR 75 and PR 162 msec. Troponin 0.04 ng/mL. Stat echocardiogram showed ejection fraction of 50%, hypokinesis of the entire myocardium, and normal LV outflow tract. The patient subsequently tolerated treatment with IV nitroglycerin infusion.

Discussion: Originally observed over a century ago, the BJR blunts inhibitory sympathetic nerve firing, leading to hypotension, bradycardia, and occasionally hypopnea. While reminiscent of a vasovagal reaction, the BJR is reported following cardiac transplantation, suggesting an intact vagal nerve is not essential to its mechanism. Classically, the BJR was understood to be chemically triggered after injection of caustic alkaloids. Subsequently, nicotine, capsaicin, and ischemia were associated with the BJR. A second mechanism postulates that severe mechanical stress of specific myocytes may cause the reaction. Animal studies support the theory that hypovolemic hypotension activates mechanical derangements stimulated by a heart contracting around an empty chamber. Clinically, the latter has occurred in profound postpartum hemorrhage, spinal anesthesia, and other settings of rapid LV emptying. In our case, SNP, a fast-acting arterial and venous vasodilator which reduces afterload, decreases ventricular filling pressure, and lowers BP, mimics physiology consistent with hypovolemic hypotension, and thus is capable of trigger the BJR. It is important to remember this rare and paradoxical reaction, which traditional physiology may otherwise struggle to explain.

References

Invasive Squamous Cell Carcinoma Masquerading as Cellulitis in the Setting of Long-term Immunosuppressive Therapy

Authors: Candace Franklin, DO, Tariq Shaheed, DO, Deborah Carlson, MD

Introduction: We present a case of unilateral lower extremity invasive squamous cell carcinoma (SCC), which was initially misdiagnosed as cellulitis. The purpose of this clinical vignette is to: 1. Bring awareness to the negative healthcare impact relative to misdiagnosis of a common inpatient problem, 2. Encourage hospitalist teams to maintain a broad differential in special patient populations including: status-post failed outpatient antibiotic therapy and those currently undergoing immunosuppressive therapy.

Case Presentation: A 57-year-old female, with a past medical history significant for end stage renal disease, prior renal transplant and current immunosuppressive therapy (prednisone and tacrolimus) presented with a 1-week history of left lower extremity erythema, edema and pain after failure of outpatient antibiotic therapy for cellulitis.

On initial examination, the patient was afebrile with all vital signs within normal limits. Physical exam was pertinent for abnormalities involving the left lower extremity (see graphic). Initial labs revealed: an elevated creatinine (3.3 mg/dL), a white blood cell count of 9.4 x 10^9/L and a normal lactic acid (1.1 mg/dL). After initial assessment, including an unremarkable doppler ultrasound, vancomycin and piperacillin-tazobactam broad-spectrum antibiotic therapy was initiated.

Given no clinical improvement after a three-day hospital course, multiple skin biopsies were obtained, and orthopedic surgery was consulted to rule out underlying necrotizing fasciitis. Although the initial Laboratory Risk Indicator for Necrotizing Fasciitis (LRINEC) score was calculated, these factors were confounded by concomitant morbidities including end-stage renal disease and chronic immunosuppressive therapy. MRI was unremarkable however, biopsy pathology results revealed “high suspicion for poorly differentiated squamous cell carcinoma involving all margins.”

Radiation therapy was scheduled given multiple large areas involving the left lower extremity. Due to no evidence of metastatic disease on CT scanning, chemotherapy was not indicated. After a nine-day hospital course, the patient was declared stable and discharged to receive outpatient focalized radiation therapy.

Discussion: Skin and soft tissue infections are a common primary inpatient diagnosis, representing 1.8% of US hospital admissions, with annual costs approximately $5.5 billion worldwide. Importantly, misdiagnosis of cellulitis is a rising problem affecting inpatient healthcare and has the potential for a negative impact in several areas including: 1. Increased healthcare costs, 2. Improper antibiotic stewardship and subsequent development of antibiotic resistance, and 3. Increased incidence of patient complications /adverse events related to non-indicated treatment options. By presenting this clinical vignette, we aim to re-iterate the importance of maintaining a broad differential diagnosis in special patient populations, as an accurate and timely diagnosis directly correlates with lower rates of morbidity and hospital-associated complications.
NORTH CAROLINA CLINICAL VIGNETTE POSTER FINALIST - MIA D GONZALES, MD

Refractory Heparin Induced Thrombocytopenia with Cerebral Venous Sinus Thrombosis treated with a Change in Anticoagulant Alongside Steroids and IVIG: A Case Report

Authors: Mia Gonzales, MD MPH, Amrish Pipalia MD, Andrew Weil MD

Introduction: Heparin-induced thrombocytopenia (HIT) type II is caused by antibody production that bind complexes between heparin and platelet factor 4 leading to platelet consumption and thrombosis. In a small subset of cases referred to as autoimmune HIT, the antibodies activate platelets even in the absence of heparin. Refractory HIT is a type of autoimmune HIT in which thrombocytopenia persists for weeks after heparin discontinuation and carries increased risk for thrombosis and more severe thrombocytopenia. We present a case of refractory HIT with cerebral venous sinus thrombosis (CVST) that was successfully treated with a change in anticoagulant alongside steroids and a second trial of intravenous immunoglobulin (IVIG).

Case Presentation: A 46-year-old woman with a history of localized breast DCIS underwent simple mastectomy with flap reconstruction at an outside hospital. Her admission platelet count was 335,000/µL and she was discharged on prophylactic-dose low molecular weight heparin (LMWH). After 8 days of LMHW therapy, she presented to the emergency department with a worsening headache. Platelet count at this time was 12,000/µL and imaging demonstrated CVST. HIT was suspected and later confirmed with a positive heparin-induced platelet antibody ELISA screen and serotonin release assay. All heparin products were discontinued and Argatroban was initiated. Despite a therapeutic partial thromboplastin time (PTT) for 7 days, there was extension of thrombosis and no improvement in platelet count, suggesting refractory autoimmune HIT. IVIg was administered for two days with no improvement. Platelet counts continued to remain low at 14 days following LMWH discontinuation. Argatroban was then switched to bivalirudin, IVIg was re-initiated for 7 days and methylprednisolone was administered once. Platelet counts subsequently demonstrated a steady rise, reaching normal levels within 5 days. She was transitioned to warfarin and her platelet count was 355,000/µL upon discharge.

Discussion: Refractory HIT occurs rarely, although the incidence is unknown. There are less than 30 case reports of IVIg used to treat refractory HIT, all of which had a rapid response in platelet count within 1-5 days after 1-2 doses of IVIg. However, our patient showed no response to an initial 2 doses of IVIg. Two of the most common anticoagulants used in HIT are Argatroban or Bivalirudin. While most studies have shown similar efficacy and safety between the two, Argatroban carries the potential for Argatroban-induced thrombocytopenia and interference with INR. Our patient was switched from Argatroban to Bivalirudin due to worsening thrombosis and persistent thrombocytopenia after failing a trial of IVIg. Corticosteroids are not a well-studied treatment modality but were administered to this patient to hypothetically suppress ongoing HIT antibody synthesis. Alongside corticosteroids and a re-trial of IVIg, this case serves to suggest a potential benefit of changing anticoagulants in cases of autoimmune HIT, particularly in those that are initially unresponsive to IVIg.

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Lethal Liquor: A Mistaken Identify of paraquat

Authors: Grant Oakley, DO

Introduction: Paraquat is an herbicide used worldwide that is exceedingly lethal even in small amounts. In 3rd world countries it is commonly used as a suicide agent given the rapidity and certainty of mortality with ingestion. It remains in use in the agriculture in the U.S. today but can only be obtained for commercially licensed use. Mechanism for injury in mammals is superoxide production with its affects concentrating in high flow organs such as the lung, kidney, liver and heart.

Case Presentation: A 65 male with history of alcohol abuse presented about 6 hours after accidentally ingesting less than a mouthful of Gramaxone® (an herbicide which contains 30% paraquat) contained in a liquor bottle of Mad Dog 20/20®. He subsequently developed GI symptoms and promptly sought medical care. Upon admission he had developed abdominal pain and was becoming increasingly lethargic. Labs were notable for creatinine of 2.61 along with lactic acidosis of 14.6 and pH of 7.21. Nephrology was emergently consulted and CVVH was initiated approximately 10 hours after ingestion. Several hours after CVVH was started his troponin markedly increased and his EKG demonstrated STEMI. Additionally patient’s respiratory status began to decline with increasing hypoxia. Given the trajectory of the patient a palliative approach was pursued after discussion with family. Patient shortly thereafter died. Time of ingestion to death took only 24 hours.

Discussion: Paraquat is a remarkably difficult toxicity to treat when ingested in high doses due to rapid action of the reactive oxygen species (ROS) and subsequent multisystem organ failure that can quickly result. At doses of 30mg/kg or 50cc of 21% paraquat solution death can ensue, however as little as 4mg/kg can still cause respiratory and renal dysfunction. Interestingly paraquat is adequately excreted through the kidneys if renal function remains intact and is more effective than hemoperfusion in removal of the toxin. However in the setting of acute paraquat intoxication AKI arises in over 50% of patients which reduces elimination of the highly toxic substance. The importance of early hemoperfusion therapy can strongly impact patient outcomes when lethal doses are ingested, but in cases such as ours even augmented removal of paraquat was unable to stall the progressive damage of the ROS. There are several other therapies that can be used in paraquat intoxication in addition to hemoperfusion. In the immediate timeframe after ingestion activated charcoal can be used which has been found to be effective in vitro, however clinical data is still lacking and it needs to be administered within 2hrs from ingestion due to rapid systemic absorption of paraquat. Immunosuppressants and antioxidants such as N-acetylcysteine, Vitmain C&E, deferoxasmine have shown some promise but have yet to be confirmed as truly effective therapies in trials.
Hematochezia in Hemophilia, Not So Simple!

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**Introduction:** Hemophilia A is characterized by deficiency in factor VIII protein which results in spontaneous hemorrhage, easy bruising, and poor clotting activity after trauma or injury (1). Inflammatory bowel disease (IBD) specifically Crohn’s disease (CD) has a variety of clinical features with hematochezia as an occasional presenting sign (2). The purpose of this case is to describe the diagnosis of Crohn’s disease in a patient with hemophilia A.

**Case Presentation:** A 19-year-old Caucasian male with a history of hemophilia A presented to our institution with a week history of hematochezia. Past medical history includes duodenal bulb ulcer requiring endoscopic therapy due to nonsteroidal anti-inflammatory drugs (NSAID) usage six years ago and anorectal abscess requiring incision and drainage with seton placement three months ago. The patient had no family history of IBD and denied usage of NSAIDs. Physical exam demonstrated a benign abdomen with rectal exam significant for a seton and bright red blood. The patient was hemodynamically stable, mildly tachycardic, and anemic with a hemoglobin of 10.7 g/dL. Factor VIII activity was low at 44% and erythrocyte sedimentation rate (ESR) and C-Reactive Protein (CRP) were elevated to 38 mm/h and 41.2 mg/L. Flexible sigmoidoscopy to rule out seton related bleeding and esophagogastroduodenoscopy (EGD) given his PUD history were negative except for dark blood material in the colon at 60 centimeters. Colonoscopy was performed showing an inflamed and erythematicus ileal cecal valve with ulceration and scar formation. Biopsy demonstrated active ulceration with granulation tissue and crypt dropout concerning for CD. A computed tomography scan found diffuse wall thickening extending from the distal ileum to the ileocecal valve consistent with CD. The patient was started on a prednisone taper and was discharged on hospital day four. During his hospitalization, the patient required transfusion of blood and Factor VIII. At outpatient follow up the patient had resolution of symptoms and was started on infliximab and Azathioprine. Repeat ESR and CRP normalized to 11 and 3.6.

**Discussion:** In the United States three million adults are diagnosed with IBD, while Hemophilia A affects only twenty thousand adults (3,4). Etiologies of hematochezia in young patients includes Meckel diverticulum, CD, and enterocolitis whereas bleeding in hemophilia occurs from gastritis, duodenitis, Mallory-Weiss tears, or angiovascular malformations (5-7). Acute GI bleeding is a rare presenting symptom of CD (8). Fistulous CD is an uncommon presenting symptom for CD with a low incidence of fistulas as the first disease manifestation (9,10). Although this patient was not diagnosed with CD during his first episode of GI bleeding or anorectal fistula, one could speculate whether these findings represented an early manifestation of CD. This case demonstrates the rarity of Crohn's disease in hemophilic patients and the unique presentation.

**References**


From sore throat to metastatic disease: the syndrome that should not be forgotten

Authors: Tamara Saint-Surin, MD

Introduction: Lemierre’s syndrome is a rare entity characterized by thrombophlebitis and septic embolization, often preceded by the more common diagnosis of pharyngitis.

Case Presentation: A 19 year old female college student with no medical history presented to Student Health two times over the course of a few days with sore throat, fevers, and body aches, and each time was sent home with supportive measures. One week after the onset of her symptoms, she presented to Urgent Care with severe back pain and difficulty breathing then was subsequently sent to the hospital. Physical exam was notable for fever, tachycardia, tachypnea, tenderness to palpation over the right trapezius, and bibasilar lung crackles greatest at the right. Labs were notable for leukocytosis, mild anemia, thrombocytopenia, low reticulocyte count, elevated alkaline phosphatase, elevated BNP, and elevated ESR & CRP. CT chest revealed numerous bilateral pulmonary nodular opacities with a basilar predominance as well as mediastinal and hilar adenopathy. CT neck with contrast revealed an occlusive thrombus in the right facial vein with partial extension into the right internal jugular vein. Echocardiogram did not reveal valve vegetations. The clinical history, embolic phenomena on chest imaging, and thrombophlebitis were consistent with Lemierre’s syndrome. Blood cultures ultimately grew Fusobacterium necrophorum and Actinomyces meyeri. The nodular opacities worsened in the first 48 hours but then remained stable over the next several days. She experienced fevers, night sweats, and pleuritic chest pain throughout most of her hospitalization. She was treated with Unasyn then Ertapenem to complete 3 weeks of IV antibiotics, which was to be followed by 9 weeks of Augmentin. She was also given IV heparin followed by low molecular weight heparin to complete 6 weeks of anti-coagulation.

Discussion

This case illustrates polymicrobial bacteremia with Fusobacterium necrophorum, the most common bacteria associated with the rare condition that is Lemierre’s, as well as with the less commonly found Actinomyces meyeri, which is a pathogen with great propensity for dissemination and has frequent pulmonary involvement. Given the extensiveness of this patient’s disease and symptoms the decision was made to anti-coagulate and to treat with a prolonged course of antibiotics (typical duration is 4-6 weeks), however a general consensus has yet to be reached on the utility of anti-coagulation and necessary duration of antibiotics in this syndrome.

References


Loperamide: A Poor Man's Methadone

Authors: Idalys Santos-Sanchez, MD

Introduction: Loperamide is an inexpensive anti-diarrheal medication that is widely available over-the-counter. It inhibits intestinal peristalsis through calcium channel blockade and peripheral µ-opioid receptor agonism. Although it is relatively safe at therapeutic doses, it can cross the blood brain barrier at high plasma concentrations and act on central µ-opioid receptors leading to psychoactive effects. It can also lead to fatal cardiac arrhythmias by acting on cardiac sodium and potassium channels. The novelty of loperamide intoxication poses management challenges.

Case Presentation: A 51-year-old man with a history of bipolar disorder, type 2 diabetes mellitus complicated by peripheral neuropathy, and opiate use disorder previously treated with methadone presented to the Emergency Department (ED) with head and neck pain one day after a motor vehicle accident. While in the ED waiting room, he collapsed and was found to be pulseless. Cardiopulmonary resuscitation was started and return of spontaneous circulation was achieved after ninety seconds. In the ED, cardiac monitor revealed a wide complex rhythm with a prolonged QRS of 156ms and prolonged QTc of 500ms. Once alert and oriented, he reported increasing use of loperamide for the past 8-9 months for management of pain from his diabetic neuropathy. He admitted to taking 200-300 mg daily (daily maximum recommended dose is 16 mg) for 1-2 weeks prior to presentation. Poison control was consulted for loperamide intoxication, and he was started on bicarbonate and hypertonic saline infusions, and intravenous lipid emulsion therapy. He continued to decompensate with further QT prolongation. He had twenty five episodes of ventricular fibrillation and went into cardiogenic shock ultimately requiring a transvenous pacemaker and extra corporeal membrane oxygenation. He was discharged five days after admission in stable condition.

Discussion: This case illustrates the challenges of loperamide intoxication management. Even though interventions are available to stabilize the cardiac electrochemical imbalances, therapy remains largely supportive as loperamide is not dialyzable, there is no reversal agent such as naloxone, and there are no medications to enhance its elimination. As physicians are becoming increasingly cautious with opiate prescribing, loperamide may provide a new, affordable, and legal way for opiate abuse and self-treatment of opiate withdrawal. As dispensing is not monitored and it is not detectable on drug screens, it is important for physicians to recognize its potential for misuse and abuse and screen patients at high risk.
Falling through the cracks – PML as initial presentation of HIV

Authors: Lauren Vaughan MD, Sowmya Nagaraj MD

Introduction: The USPSTF and CDC both recommend one time HIV screening for individuals aged 15-65 years old with repeated screening indicated in patients at high risk. Despite the recommendations, the CDC estimates that over 150,000 people living in the US with HIV are unaware of their diagnosis. Screening for HIV is essential as prompt diagnosis and linkage to care with timely initiation of highly active antiretroviral therapy (HAART) early in the disease progression lower the risk of development of AIDS and fatal AIDS defining illnesses. Additionally, knowledge of HIV status has been shown to encourage people to adopt safer lifestyle behaviors suggesting that testing itself is an effective intervention in preventing the transmission of the virus. All clinical encounters ranging from outpatient care to emergency department visits are an opportunity for HIV screening and early detection.

Case Presentation: 33-year-old female with medical history of hypertension and gestational diabetes presented with one-week history of new onset left-sided weakness, facial droop, slurred speech, ataxia and headache. CT head was negative and brain MRI revealed a right cerebral hemispheric mass. Lab studies revealed positive HIV antibody, negative p24 antigen, and CD4 count of 50, indicative of AIDS. The patient was unaware of her HIV status despite multiple healthcare encounters throughout the years. These included primary, ED, and OB/GYN visits plus receiving prenatal care for her three children. Patient underwent brain biopsy with pathology showing extensive inflammatory cell infiltrates and intranuclear inclusions, consistent with progressive multifocal leukoencephalopathy (PML). CSF analysis was JC virus positive, confirming the diagnosis of PML, an AIDS-defining disease. She was started on HAART while inpatient and discharged to inpatient rehabilitation.

Discussion: Often patients newly diagnosed with HIV have had multiple health care encounters prior to their diagnosis, each representing a true missed opportunity as early detection allows for access to treatment and decreased morbidity and mortality. These missed opportunities lead to the high prevalence of “late presenters,” or patients diagnosed at advanced stages with CD4 count below 350 or presenting with an AIDS-defining illness. Medical providers must take into account the risk factors and clinical indicators and subsequently lower their threshold for HIV testing as to prevent late presentation. Despite established screening recommendations and access to free HIV testing services being readily available throughout the US, thousands of individuals go untested and undiagnosed. The question then becomes determining and combating the barriers to HIV testing.

Spontaneous HIT following Total Knee Arthroplasty

Authors: Monu, Minnu MD; Potluri, Rajendra MD

Introduction: Spontaneous HIT is a rare syndrome which offers a similar serological presentation as HIT without any preceding Heparin exposure. Occurrences of spontaneous HIT are fairly rare and its etiology is not well established and needs further research.
Case Presentation: A 63-year-old female presented on 8/1/2018 with symptoms of alexia and aphasia of 3 day duration, following which she was found to have a left temporal intraparenchymal hemorrhage with edema and a new onset thrombocytopenia.

She recently had undergone a right total knee arthroplasty on 7/12/2018 and was placed on Apixaban for antithrombotic prophylaxis for a period of 11 days. There was no documented intra/postoperative heparin exposure or with other heparin related products. There was also no previous history of thrombocytopenia, infection, or other recent acute illness.

Based on her acute onset stroke, a CTA of the head was performed which revealed a thrombus in the left sigmoid sinus, with bleeding in the left temporal lobe. Further a bilateral Lower extremity ultrasound showed Deep vein thrombosis. Her platelet count dropped from 267(7/12/2018) to 53(8/1/2018).

Based on her symptoms and recent literature showing incidence of spontaneous HIT following Total knee arthroplasty we suspected the same and patient was started on Argatroban at a low dose 0.1 mcg/kg/hr, with a target APTT around 1.5 upper normal in 50 Sec. To confirm the diagnosis HIT antibody optimal density test and Serotonin release assay was ordered. Her HIT Optical Density test came back positive at 2.6, and Serotonin release assay was positive (92% release at 0.1 UFH and 97% release at 0.5 UFH) and helped confirm our diagnosis of spontaneous HIT.

Discussion: We report a rare spontaneous HIT syndrome following a total knee arthroplasty. The exact pathophysiology of spontaneous HIT remains unclear but is thought to be due to the interaction of negative polyanions such as nucleic acids and chondroitin sulphate with PF4. The formation of IgG : PF4 : polyanion complexes leads to platelet activation via Fcg receptors, which in turn increases predisposition to venous and occasionally arterial thrombosis.

There have been 11 cases of spontaneous HIT reported in literature and of these 9 have occurred post a Total Knee Arthroplasty. Spontaneous HIT is not widely known and harder to diagnose however as the patient presents with HIT like symptoms without any previous exposure to heparin. The syndrome can cause both thrombocytopenia as well as thrombosis and can thus be fatal if not promptly diagnosed. Physicians therefore need to be aware of this syndrome and must consider it in their differential diagnosis especially if the patient has recently undergone a knee/shoulder arthroplasty.

References

A case of nodal anthracosis presenting as PET-positive mediastinal and hilar lymphadenopathies

Authors: Jafar Alzubi MD, Ahmad Jabri MD, Hussein Hussein MD

Introduction: Anthracosis is a form of pneumoconiosis seen most commonly in coal workers, although other environmental factors including cigarette smoke, air pollution, and biomass fuels used for cooking and home heating are also known causes. The objective of this report is to describe a patient with FDG PET/CT-positive mediastinal and hilar lymph nodes, confirmed to be primary nodal anthracosis by endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA).

Case Presentation: A 78-year-old Nepali female non-smoker who was referred to the pulmonary clinic for evaluation of a 1.1 cm left upper lobe lung nodule discovered during work up for her dyspnea on exertion and heart failure exacerbation. Past medical history included atrial fibrillation, latent tuberculosis, pulmonary emphysema and congestive heart failure.

A PET-CT scan of the chest demonstrated normal metabolic activity and old scarring in the left upper lobe, but revealed FDG avid bilateral hilar and mediastinal lymph nodes, worrisome for malignancy. The patient underwent EBUS-TBNA of the subcarinal lymph node located at station (7) and left paratracheal lymph node located at station (4L), which subsequently revealed black-colored material exiting the lymph nodes at the sites of needle insertion. Fine needle aspiration was negative for acid fast bacilli (AFB), fungi and malignancy. However, on microscopic examination, there was abundant anthracotic pigment consistent with the diagnosis of nodal anthracosis. Later, it was discovered that this patient had significant exposure to biomass fuels used for cooking in Nepal as well as exposure to outdoor air pollution.

Discussion: The patient described here had PET-positive mediastinal and hilar lymphadenopathies, confirmed on microscopic examination to be nodal anthracosis. Individuals with anthracosis may develop both physical and radiologic abnormalities of the lung presenting as chronic obstructive or fibrotic lung disease due to chronic exposure to smoke and particulates. Extrapulmonary anthracosis is rare, and can present as a mediastinal mass, mediastinal or hilar lymphadenopathy with PET-positivity, as with this patient, mimicking infectious conditions, granulomatous diseases and malignancies. Thus, accurate lymph node sampling with EBUS-TBNA is necessary to establish a diagnosis of nodal anthracosis.

Anthracosis should be considered in the differential diagnosis of PET-positive hilar or mediastinal lymphadenopathy, particularly in the presence of black-colored deposits observed along the airway during lymph node sampling,. Intentional questioning on occupational history, significant exposure to outdoor pollution and/or biomass fuels for cooking or heating in such cases may expedite course to diagnosis and treatment.
Necrotizing Autoimmune Myositis; A Rare Complication of Statin Therapy

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Introduction: Necrotizing autoimmune myositis (NAM) is a newly described clinical entity that can occur following years of statin use. It is characterized by progressive weakness, elevated creatinine kinase (CK) levels, myonecrosis, and anti-HMGCoA antibodies. The recent discovery of anti-HMGCoA antibodies (2010) has allowed for both the diagnosis and prognostication of patients with NAM, with higher antibody levels being associated with a more severe clinical course. Although a rare condition with an estimated 2 cases per million reported per year, the wide use of statins and the severity of NAM requires all physicians to be aware of this disease.

Case Presentation: A 68 year old male with a history of type II diabetes, hypertension, and hyperlipidemia presented with a 2-week history of diffuse muscle aches, proximal muscle weakness, and difficulty ambulating. He also noted a 20 pound weight loss and darkening of his urine during this period. His medications were metformin, sitagliptin, lisinopril, amlodipine, and atorvastatin, which he had been taking for 3 years.

Labs on admission showed: CK: 10,197 U/L, ALT 674 U/L, AST 429 U/L, GGT 24 U/L, ESR 52 mm/hr, CRP 4 mg/dL and creatinine of 0.47 mg/dL (baseline 0.7-0.9). He was admitted for further management of presumed myositis.

MRI of the thighs showed diffuse edema. ANA and myositis panel, including Anti-Jo-1, anti-Ro-52, anti-SRP, and anti-Mi-2 were negative. Anti-HMGCoA was sent and 9 days after presentation it resulted in a strongly positive titer (>200). He was started on prednisone and azathioprine soon after admission, however developed severe dysphagia and respiratory compromise. IVIG was initiated, the patient improved, and was discharged to a rehab facility.

He was re-admitted 2 weeks later with worsening dyspnea resulting in PEA arrest with successful resuscitation. Rituximab was then added. He was discharged to a rehab facility on a ventilator with methylprednisolone, AZT, and weekly IVIG. 6 months after discharge he has been weaned off the ventilator and is improving with physical therapy.

Discussion: Statin-associated myalgias are a common complaint in the primary care setting, with an incidence between 9-20% of statin users. Severe myopathies are much more rare, reported in less than .1% of all statin users. Both myalgias and myopathies improve with withdrawal of statin therapy and supportive care. By contrast, NAM is a potentially fatal autoimmune disease that follows an aggressive clinical course if not quickly recognized. NAM can be distinguished from statin-associated myalgias and myopathies by: the degree CK elevation (often >10,000 IU/L), the presence of anti-HMGCoA antibodies, and the lack of improvement after discontinuation of statin therapy.

As seen in this case, NAM can be a devastating complication of statin use. Early detection, discontinuation of statin therapy, and prompt treatment with immnosuppressive agents may improve overall outcomes.

References

Uncontrolled Diabetes Mellitus: A Rare Cause of Central Pontine Myelinolysis

Authors: Nicholas Amata MD; Kim Jordan MD FACP

Introduction: Central Pontine Myelinolysis (CPM) is usually attributed to rapid correction of electrolytes in the setting of chronic hyponatremia, typically with sodium levels less than 120 mmol/L. First described in 1959 in patients who were malnourished or had alcoholism, other etiologies for CPM are reported; however, physicians may not consider the diagnosis in the setting of normal serum sodium levels in patients presenting with neurologic abnormalities. Diagnosis is often made only after confirmatory neuroimaging documentation of hyperintensities in the pons region. We describe CPM in the setting of chronic poorly controlled diabetes mellitus.

Case Presentation: A 50-year-old male with hypertension, diabetes mellitus, and alcohol abuse (sober for 2 months) presented to the hospital following a witnessed tonic-clonic seizure. The patient reportedly “was well” until the day of presentation, when his significant other noted gait unsteadiness and need to hold onto walls for balance. On examination, the patient had decreased attention, mixed aphasia, and slurred speech without other neurologic deficits. His mental status was rated 3 on The National Institutes of Health Stroke Scale. His sodium was 132 mmol/L, potassium 5.0 mmol/L, alcohol undetectable, and glucose 579 mg/dL. Sodium corrected for hyperglycemia was 140mmol/L and hemoglobin A1c was 17.9%. Brain computed tomography showed no acute intracranial abnormality, and EEG showed no epileptiform activity. However, brain MRI showed hyperintense signal on T2 and FLAIR centrally in the pons and restricted diffusion involving the central portion of the pons, with abnormality measuring 1.5cm x 2.4cm; a diagnosis of CPM was made. He improved with blood glucose normalization, and by hospital day 2 had no further neurologic symptoms. The patient subsequently reported that he could not afford his prescribed insulin regimen, had been borrowing friends’ insulin, and filling his prescriptions inconsistently. He was started on a more affordable insulin regimen and discharged with close outpatient follow-up by his primary care physician and neurology.

Discussion: CPM is a rare complication of diabetes mellitus, despite the pronounced fluid and electrolyte shifts caused by hyperglycemia and insulin administration. The majority of case reports describing osmotic demyelination in association with diabetes have occurred in patients with concomitant ketoacidosis, or concurrent sodium abnormalities, or following treatment of the hyperosmolar state. Importantly, the clinical presentation of CPM is variable, ranging from altered consciousness, dysarthria, to flaccid quadriplegia. Diagnosis is made with demonstration of abnormalities in the pons on imaging, though radiological lesions extending beyond the pons are recently reported. Prognosis is not dependent on the burden of radiologic abnormalities, and outcomes vary from complete recovery to death. Treatment is largely supportive and aimed at underlying etiology.
Pasteurella multocida endocarditis in a patient with congenital asplenia

Authors: Courtney Certain DO, Kim Jordan MD

Introduction: Pasteurella multocida, a small gram-negative coccobacilli, is the most common isolate found in infected cat scratch/bite injuries (75%). Infection generally occurs in the setting of close contact (bed sharing, licking, biting and scratching) and most resolve without complication. The most common manifestation of P multocida infection is cellulitis and soft tissue infection. Oral and respiratory infection can occur, particularly if underlying chronic pulmonary disease. Serious invasive infection, including endocarditis from P. multocida is rare.

Case Presentation: A 67-year-old female with systemic lupus erythematosus (SLE), on long term plaquenil therapy, and congenital asplenia presented with low back pain, nausea and vomiting. Physical exam showed purpuric/petechial lesions of pus of her fingers and autoamputation changes to the right great toe. Initial cefepime therapy for presumed urinary tract sepsis was started, but was changed to ceftriaxone when blood cultures resulted positive for Pasteurella multocida. Further history revealed that the patient had recently adopted a feral cat and sustained a scratch to her right leg approximately one week before presentation. Initial surface echo was negative for vegetation but on day 3 of hospitalization a new murmur was noted. Repeat echo then revealed 2.1x1.1cm posterior annulus mitral valve vegetation without abscess and new reduced ejection fraction. Despite therapy with ceftriaxone, the patient developed severe thrombocytopenia (platelet count of 33,000), quadratus lumborum and psoas myositis, multilevel discitis, epidural abscess, innumerable septic cranial infarcts, ischemic limb with concern for necrotizing fasciitis, and multiorgan damage. In the setting of the widespread infection, further work up and intervention (including epidural abscess drainage, revascularization of ischemic limb and valvuloplasty) were not possible. Despite aggressive therapy and a multidisciplinary team approach, she failed to respond and on hospital day 14, the patient opted for comfort measures only. She expired 22 days after presentation.

Discussion: This case demonstrates a pet-related fulminant, catastrophic infection in an immunocompromised adult. Severe sepsis and endocarditis from Pasteurella multocida infections are rare, reportedly occurring in 7.8% - 11% of infections. Generally, infection occurs in older adults with underlying chronic disease, including diabetes mellitus, chronic renal or liver disease, and/or other immunocompromising states (our patient had known SLE and was asplenic). Treatment for P. multocida blood and deep tissue infections include high-dose IV penicillin or alternatively, ampicillin-sulbactam, piperacillin-tazobactam or broad spectrum cephalosporins (including ceftriaxone) for beta-lactamase producing infections. Early detection of disease progression in high-risk populations is tantamount to treatment success; however, mortality rates are high when P. multocida bacteremia occurs, ranging from 7-31%.

References

A Case of Embolic Stroke Originating in a Pulmonary Arteriovenous Malformation.

Authors: Johnny Chahine, MD; Bicky Thapa, MD; Raunak Nair, MD; Oscar Perez Gomez, MD; Anjli Maroo, MD. Cleveland Clinic, Fairview Hospital, Cleveland, Ohio, USA.

Introduction: Ischemic strokes, as opposed to hemorrhagic strokes, constitute 80% of cerebrovascular accidents. The source of emboli might be cardiac, vascular, or unknown. We hereby present a rare source of embolic stroke, a pulmonary arteriovenous malformation (PAVM), suspected on contrast echocardiography.

Case Presentation: A 64-year-old female with a past medical history of major depression, bipolar disorder, hypertension, hyperlipidemia and chronic obstructive pulmonary disease presented to the emergency department after a fall that was preceded by rotational vertigo. She also had left upper extremity numbness and weakness. She denied loss of consciousness, difficulties with speech, and other areas of weakness. She had had multiple similar episodes in the preceding two weeks. She endorsed a strong family history of stroke. On arrival to the emergency department, her vertigo, weakness, and numbness resolved. Her vital signs were stable. She had a dysconjugate gaze but an otherwise normal neurological exam. Magnetic resonance imaging of the brain showed scattered small bilateral acute infarcts involving multiple vascular territories without significant mass effect or hemorrhagic conversion. A computed tomography angiography of the head and neck showed left cervical internal carotid artery with a small ulcerative plaque. She was treated with simvastatin, aspirin, and clopidogrel. Transthoracic echocardiogram with agitated saline contrast showed delayed opacification of the left atrium after contrast administration, suggestive of intrapulmonary shunting. Transesophageal contrast echocardiogram confirmed significant delayed right to left shunting, suspicious for pulmonary arteriovenous malformation. A large pulmonary arteriovenous malformation was visualized on computed tomography of the chest, and the patient was scheduled for coil embolization of the PAVM.

Discussion: The prevalence of PAVMs is estimated to be around 0.02%. They are more common in women and are mostly congenital. More than half of PAVMs remain asymptomatic; the remainder can present with respiratory manifestations (shortness of breath, chest pain, hemoptysis and lung nodules), bleeding, and neurologic symptoms due to brain abscesses or strokes. Neurologic symptoms are believed to be due to a paradoxical embolus arising from the deep veins. In our patient, the involvement of multiple cerebral vascular territories is suggestive of a cardioembolic stroke. Although a small ulcerated plaque was seen in the left internal carotid artery, the multiple vascular territories affected by the stroke pointed more to a central embolic source. Also, the presence of left-sided weakness and numbness was not congruent with left internal carotid artery pathology. PAVM is an uncommon cause of embolic stroke, but it is amenable to treatment with percutaneous coil embolization. Contrast echocardiography was a crucial first step in establishing the diagnosis of PAVM. In patients with strokes that have a high probability of a central embolic source, contrast echocardiography should be performed as part of the initial diagnostic evaluation.
Hypertension, A Headache in Disguise

Authors: Logan Dalal MD, Kim Jordan MD FACP

Introduction: Hypertension is a pervasive condition, contributing to neurological, cardiovascular, and renal disease. Comorbid conditions including obesity, diabetes, smoking, and alcohol use increase the risk of hypertension, with only 52.5% of the affected population meeting AHA guidelines for adequate control. Approximately 5-10% of cases of hypertension are considered secondary in etiology, with potentially reversible causes. This case illustrates a rare case of renal artery compression and subsequent renovascular hypertension secondary to progression of Non-Hodgkin’s Lymphoma (NHL).

Case Presentation: A 48-year-old female presented with a ten-day history of left-sided headache with central vision loss and facial pain. Her blood pressure (BP) was elevated to 210/104, prompting hospitalization. She had known grade 2 follicular NHL under surveillance, and surgical hypothyroidism following resection of papillary thyroid carcinoma, but denied prior diagnosis of hypertension. On arrival, BP was 234/131, but improved to 177/90 with IV hydralazine. Computed tomography (CT) angiography of her head and neck was normal with no acute process. Thyroid studies and urine toxicology screen were negative, but serum creatinine was elevated at 1.09mg/dl (above baseline) and hypokalemia of 3.2 was noted. Urine 24-hour metanephrine, serum renin, and aldosterone levels were normal. Carvedilol and lisinopril were started. Renal duplex ultrasound was negative for high flow gradient or stenosis of the renal arteries, but left hydronephrosis was present. Subsequent abdominal CT showed increased retroperitoneal adenopathy and a necrotic 5.4 cm by 4.5 cm mass that narrowed and enveloped the left renal artery and vein, and obstructed the left renal collecting system. Core biopsy of the mass and flow cytometry were consistent with monoclonal B-Cells with CD10 expression. Her lymphoma was restaged to 3A, Grade 1-2 and chemotherapy was initiated. Her BP at discharge was well-controlled. At one month follow-up, after directed chemotherapy, the patient’s hypertension had resolved, and antihypertensive medications were discontinued. Final pathology showed evidence of “double-hit” BCL-2 and MYC rearrangements, and her chemotherapy regimen was modified to EPOCH with rituximab. At four months follow-up, CT scan showed a decrease in left retroperitoneal mass size and resolution of obstructive uropathy. Her BP remained normal off all anti-hypertensive medications.

Discussion: Renovascular hypertension is most commonly associated with renal artery stenosis in the setting of fibromuscular dysplasia, embolic disease or atherosclerotic changes. Extrinsic compression due to tumor burden is uncommon. Follicular NHL typically presents at diagnosis with painless lymphadenopathy, or imaging suggestive of new lymphadenopathy; between 2 and 14% percent of patients present with renal failure due to ureteral obstruction. On initial diagnosis, asymptomatic patients undergo watchful waiting. However, some are at risk for aggressive transformation with BCL-2 and MYC overexpression. Acute onset hypertension, in patients with low comorbidities should prompt immediate and swift evaluation for secondary factors.
A Rare Case of Myocarditis Secondary to an Occult Malignancy

Authors: Aneesh Dhorepatil MD, Amit Arbune MD, Khalil Murad MD MS, Ashish Aneja MD

Introduction: Eosinophilic myocarditis (EM) is a relatively rare condition that may result from hypereosinophilic syndromes (HES). It manifests clinically with acute heart failure, cardiogenic shock, cardiac arrhythmia, or cardio-embolic events. Renal cell carcinoma (RCC) is a rare cause of HES. To the best of our knowledge, there has been no reported case of EM secondary to HES in a patient with renal cell carcinoma. In this report, we present a case of EM in a patient with HES secondary to RCC.

Case Presentation: This is a 61-year-old man with a history of hypertension and asthma who presented to the emergency department with a 3-week history of progressive pleuritic and sharp chest and back pain for over 3 weeks, and nearly 50lb weight loss over the preceding 4 months. Laboratory investigation revealed markedly elevated absolute eosinophil count (AEC) of 27.3 x 10³/uL and elevated troponin-I level of 35.3ug/L. Electrocardiogram demonstrated ST and T wave changes suggestive of myocardial ischemia. Transthoracic echocardiogram revealed globally reduced left ventricular (LV) systolic function with an ejection fraction of 40%. Coronary angiography revealed no obstructive coronary disease. Cardiac Magnetic Resonance revealed increased subendocardial signal on T2 weighted images in the LV myocardium. Delayed post-gadolinium imaging showed diffuse subendocardial hyper-enhancement within the LV, most consistent with the hypereosinophilic (HES) state. Based on the typical clinical presentation and findings on imaging studies, endomyocardial biopsy was deferred and the diagnosis of EM was made.

Investigations for the cause of the HES including a bone marrow biopsy and flow-cytometry, and serological testing were negative for hematological malignancies, infectious, or autoimmune causes. Upon investigating the cause of his pleuritic chest and back pain, a Chest Computed Tomography revealed a 4.1?cm enhancing mass in the upper pole of the right kidney, highly suspicious for RCC, which was confirmed by renal biopsy showing oncocytic variant of papillary RCC (pT1a pNX).

In addition to guideline-directed medical therapy for systolic heart failure, the patient was started on high dose corticosteroid therapy which resulted in a rapid drop in AEC to 0.087 x10³/uL. Oral anticoagulation was also started to reduce risk of LV thrombosis. The patient subsequently underwent surgical resection of the renal mass and is being tapered off corticosteroids with sustained resolution of hypereosinophilia.

Discussion: EM is a serious complication of HES, which can be associated with underlying malignancy. The clinical presentation can range from cardio-embolic events and arrhythmias to cardiogenic shock and acute fulminant myocarditis.

The acute management of HES with EM revolves around early administration high dose corticosteroids and management of heart failure. Optimal duration of corticosteroid treatment is an avenue requiring additional research.

Thrombogenicity and need for anticoagulation in HES with EM is controversial. Endoventricular thrombi have been observed in 12.3% of patients with histologically proven EM and 28.6% in HES with EM. These findings seem to suggest a role for anticoagulation in the acute phase of EM. Further studies and trials will be needed to assess risk vs benefit of anticoagulation.

References

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A rare colon tumor

Authors: Mohamed Eisa MD, Birju Shah DO, Hope Hastings MD, Dan X. Cai MD, Dalbir S Sandhu MD

Introduction: Granular cell tumors (GCTs), also known as Abrikossoff tumors, are soft-tissue neoplasms with probable origin from Schwann cells that are very rarely found in colon. Here we report case of a colon GCT found on screening colonoscopy. In addition, the current available literature on this rare tumor is also reviewed (Table 1).

Case Presentation: A 51-year-old female was found to have a 4-millimeter polyp in the ascending colon on screening colonoscopy (Figure 1). There was no evidence of synchronous lesions. The polyp was removed en-bloc using cold snare resection without any complications. Histological evaluation revealed granular eosinophilic cytoplasm with nuclei that were small and uniform in appearance consistent with a GCT (Figure 1). On immunohistochemistry, Inhibin A and S100 were strongly and diffusely positive in the neoplastic cells confirming the Schwannian origin of the lesion. Due to the small size and essentially benign nature on histopathology, we plan to repeat colonoscopy in 3 years.

Discussion: GCTs are rarely found in the intestinal tract with esophagus being the commonest site followed by the colon, stomach, small intestine, and anal canal. Most cases are found incidentally. They present commonly as small, sessile, round, submucosal nodules covered by normal mucosa. Commonly solitary lesions, up to 10 percent can present as two or more lesions at diagnosis. The diagnosis is usually made by histopathology. Neoplastic cells are plump, histiocyte-like, and bland-appearing with abundant granular eosinophilic cytoplasm containing acidophilic, periodic acid-Schiff (PAS) positive, diastase-resistant granules with small, uniform nuclei, in which mitotic figures are absent and uniform expression of neural markers, including S100 protein or neuron-specific enolase is noted. Advanced techniques have made endoscopic removal of these lesions a viable treatment option. Given the reported recurrence of GCTs, follow up is mandatory to identify recurrence and to assess the effectiveness and oncologic safety of endoscopic resection. Factors that predict higher malignant potential of GCTs are size (especially if it is more than 4 cm), presence of symptoms, depth of invasion, and site of primary tumor. For this reason inability to completely remove the lesion endoscopically or persistent positive margins mandate segmental resection. Continued reporting of such cases can help reach a management consensus and improve our understanding of this rare disease entity.
Spontaneous Isolated Superior Mesenteric Artery Dissection Associated With Cocaine Abuse: A Case Report


Introduction: Spontaneous isolated superior mesenteric artery dissection (SISMAD) is a rare but potentially fatal arterial disease and is defined as dissection of the superior mesenteric artery (SMA) without involvement of the aorta, celiac, inferior mesenteric, or renal artery (1, 2, 3). Risk factors have been suggested (arteriopathy, tobacco, atherosclerosis, alcohol, pregnancy) but cocaine’s relationship with SISMAD is yet to be characterized. We present a case of SISMAD associated with cocaine abuse treated with conservative management.

Case Presentation: A 38-year-old African American male with a history of hypertension, major depression, diverticulosis, alcohol, cocaine, and tobacco use presented with recurrent abdominal pain that was crampy and intermittent for the previous couple of days. He endorsed recent use of cocaine. Blood pressure was 190/120 mm Hg. On examination, his abdomen was soft with diffuse abdominal tenderness. A computed tomography (CT) angiogram revealed a SISMAD within the proximal-to-mid SMA with thrombosis of proximal false lumen and no obvious re-entry point. He was successfully treated with conservative management with resolution of his abdominal pain and hypertensive urgency and was discharged home with outpatient surveillance CT angiograms.

Discussion: SISMAD is a rare disease with <1000 cases reported (3, 4). Although its presentation can be variable, the severity of the pain has been shown to correlate with the length of dissection (5). CT angiography is considered the gold diagnostic standard(6). Among multiple classification systems, Yun’s classification has become the most widely used due to its simplicity (5). Using Yun's classification, our patient’s SISMAD is classified as a type 2a/2b SISMAD; Yun type 2 SISMADs are found to be more likely to undergo complete remodeling (8).

Arterial wall dysfunction and increased shear stress are suggested to play role in the pathogenesis of SISMAD and both are also involved in cocaine-mediated arteriopathy (11, 12). In aortic dissection, cocaine causes impairment of elasticity and apoptosis of vascular smooth muscle cells (VSMCs) (17, 18). In SISMAD, elastic fiber fragmentation and loss of VSMCs are also seen histologically (3). Also, cocaine’s increased sympathetic drive increases shear stress especially at the ligamentum arteriosum which is a point of fixation (14). Similarly, flow dynamic studies have shown that the SMA’s transition from a retro-pancreatic position into a mobile mesenteric position leads to increased shear stress (11, 12). With the aforementioned mechanisms, it is postulated that our patient’s acute and chronic cocaine abuse may have resulted in gradual SMA wall dysfunction and elevated shear stress forming a nidus for subsequent dissection.

SISMAD treatment includes conservative management, endovascular therapy, or open surgery (3). Conservative management remains the most common modality (19). To the best of our knowledge, there are no cases of cocaine-related SISMAD to recommend specific treatment strategies. Our report emphasizes the need to consider cocaine abuse in SISMAD pathophysiology and treatment algorithms in future studies.

References


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Introduction: Pubic symphysis septic arthritis (PSSA) is rare. To our knowledge, septic arthritis of pubic symphysis caused by Pasteurella multocida has not been previously reported. We report the first case of PSSA caused by P. multocida and review the literature of PSSA and P. multocida septic arthritis.

Case Presentation: A 79 year old Caucasian woman presented to the Emergency Department with a 3-day history of progressively severe left groin pain and swelling. Pain was exacerbated by lying on her left side and improved with oral naproxen. Patient described an unintentional 20-lb weight loss over the prior nine months. Past medical history: hypertension, hyperlipidemia and spinal stenosis. Past surgical history: remote right knee arthroscopy, remote uterine dilatation and curettage. There was no history of alcoholism or substance abuse. Examination: heart rate = 83 beats/min., respiratory rate = 16 breaths/min., blood pressure = 106/58 mmHg, temperature = 98.2°F. In the left groin/suprapubic area there was a semi-fluctuant, tender mass approximately 6cm x 2cm dimensions extending from mid-pubic symphysis area to the left femoral crease. There was no erythema or adenopathy. CT abdomen/pelvis: multi-loculated mass in the area of the pubic symphysis measuring 7.6 cm x 2.1 cm x 5.5 cm with fluid extending posteriorly behind the pubic symphysis. WBC = 12,3000/µL [3,700-10,700/ µL], CRP = 207.8 mg/L [0.0 – 2.6 mg/L], ESR = 83 mm/h [0-20 mm/h]. Ultrasound-guided aspiration of loculated mass yielded 6 ml of purulent fluid. Gram-stained smear of aspirate showed PMNs and Gram-negative bacilli. Intravenous piperacillin/tazobactam was begun. On day 2 the patient underwent operative drainage of subcutaneous/subfascial fluid loculations. Cultures of needle aspirate and surgical specimens showed a pure growth of Pasteurella multocida. 2 pre-antibiotic blood cultures remained sterile. Further history revealed that the patient has one pet cat. The cat has only one tooth remaining and the patient has not been bitten or scratched by the cat. On hospital day 5 the patient required repeat surgical drainage for persistent 1.3 cm x 1.6 cm painful mass centered around the pubic symphysis. Antimicrobial therapy consisted of IV piperacillin/tazobactam (3 days), IV ampicillin/sulbactam (10 days) and oral amoxillin/clavulanate (5 days). The patient returned to normal with no recurrence in 3 months’ follow-up.

Discussion: PSSA is rare. No cases were reported in 6 large published series of septic arthritis (738 cases). Septic arthritis caused by Pasteurella multocida is not unusual but searches in PubMed and Google Scholar databases revealed no cases of P. multocida PSSA and only one case of P. multocida septic arthritis of any axial joint (sternoclavicular). We review PSSA and septic arthritis caused by P. multocida
Pasteurella Multocida Meningitis After a Non-Bite Animal Exposure

Authors: Sami Ghazaleh, MD; Emad Abu-Sitta, MD; Ragheb Assaly, MD; Caytlin Deering, DO, University of Toledo Medical Center, Toledo, Ohio

Introduction: Pasteurella multocida is known for causing skin and soft tissue infections following animal bites. Invasive Pasteurella infections have been reported following non-bite animal contacts, especially in patients with impaired host defenses. We describe a case of Pasteurella multocida meningitis with a complicated hospital course.

Case Presentation: A 65-year-old Caucasian female patient presented to the hospital with a four-day history of altered mental status associated with fever, nausea, and generalized weakness. Before admission, she was found wandering inside her house confused, naked, and covered with cat feces. Past medical history was significant for chronic obstructive lung disease and alcoholic liver disease. Social history was positive for smoking tobacco and consuming alcohol daily. She lived in a trailer with her boyfriend and twenty-five domestic and stray cats.

On physical exam, she was lethargic and disoriented. Vital signs demonstrated a temperature of 36.5°C, blood pressure of 99/60 mmHg, heart rate of 114 beats per minute, respiratory rate of 20 breaths per minute, and O2 saturation of 97% on room air. Neurological exam showed no focal neurological deficit. The rest of her physical exam including extensive skin examination was unremarkable with no scratches or bites.

Laboratory findings were significant for WBC of 30.6 x 10^9/L, lactate of 2.7 mmol/L, and procalcitonin of 1.51 ng/mL. Her liver function tests were within normal range, and her urine toxicology screen was negative. CFS analysis demonstrated WBC of 5817/mm3, protein of 1188 mg/mL, and glucose of 40 mg/dL. Blood and CSF cultures grew Pasteurella multocida. CT of the head without contrast showed no acute abnormalities. MRI of the brain demonstrated hydrocephalus and proteinaceous fluid suggestive of pus inside the lateral ventricles.

The patient was admitted to the intensive care unit as her mental status deteriorated and she was intubated for airway protection. She was started on IV ceftriaxone and initially improved. Her hospital course was later complicated with ventilator-associated pneumonia secondary to Pseudomonas aeruginosa, acute heart failure with low ejection fraction, and left ventricular thrombus. Ceftriaxone was modified to cefepime, and anticoagulation was started with IV heparin infusion. After 10 days of effective antimicrobial therapy, the patient’s mental status did not improve. Therapy was escalated and intraventricular access was obtained. Intraventricular gentamicin was given for a total of 7 days. Her mental status slowly improved and she was later extubated. Unfortunately, her mental status did not return to baseline at the time of hospital discharge.

Discussion: This case illustrates how Pasteurella multocida can be transmitted from animals to humans even in the absence of bites or scratches. The organism is a rare cause of meningitis but should be considered in patients with impaired host defenses (e.g. liver disease) and history of non-bite animal exposure.

References

ACUTE PANCREATITIS IN SICKLE CELL VASO-OCCCLUSIVE CRISIS- A RARE PRESENTATION OF A COMMON DISEASE

Authors: We'am Hussain M.D., Nneoma Onuorah M.B.B.S., Immaculate Foy M.D., Marc Raslich M.D.

Introduction: Sickle cell disease (SCD) is a genetic autosomal recessive disorder resulting from a mutation of Hemoglobin S, characterized by vaso-occlusive crisis (VOC) and hemolytic anemia. Vaso-occlusion is caused by sickle-shaped red blood cells (RBCs) that obstruct micro vessels, ultimately resulting in mesenteric ischemia, pain and necrosis\(^1\). Acute abdominal pain is a common presentation of VOC, which could involve any of several abdominal organs. However, acute ischemic pancreatitis is a very rare complication of VOC\(^2\).

Case Presentation: A 19-year-old woman with homozygous sickle cell disease, hemoglobin SS, and a history of cholecystectomy, and no history of alcohol abuse or hypercholesterolemia, presented with nausea, vomiting, abdominal pain, and bilateral hip, knee, and ankle joint pain. She is well known to our treatment team as medical history includes multiple hospitalizations for abdominal pain from VOC. Physical exam was remarkable for epigastric tenderness radiating backwards, nausea and vomiting. The pain was similar to previous painful VOC in the past. Patient was afebrile, with no leukocytosis and lipase was elevated at 1468U/L. Computed tomography of Abdomen and Pelvis was normal. Symptoms resolved with intravenous fluid (IV) hydration and pain medications, and diet was advanced from clear fluid diet to regular diet by day 5 of admission.

Discussion: Abdominal pain is a common presentation of sickle cell VOC, with involvement of any abdominal organ as a result of capillary engorgement, sickling of RBCs, hypercoagulability and stasis in the vasa vasorum of larger vessels\(^3\). Common etiologies in these patients include acute splenic sequestration crisis, splenic infarction, ischemic bowel, biliary colic, acute cholecystitis, opioid induced constipation, renal papillary necrosis, hepatic crisis, Urinary tract infection, hepatitis and liver abscesses\(^5\). Acute pancreatitis rarely occurs in VOC sickle cell crises\(^6\). Akingbola et al in a 2011 small prospective study showed that 70% of acute abdomen in VOC involved the epigastric region with similar presentation to acute pancreatitis\(^7\). When it occurs, it may be due to biliary obstruction, or microvessel occlusion causing ischemia\(^6\). Despite acute abdominal pain being a relatively common complaint in patients with VOC, life-threatening complications can occur if micro vessel occlusive induced pancreatitis is missed, since it is clinically indistinguishable from acute abdominal pain from sickle cell VOC and initial diagnosis is based on clinical suspicion. Our patient met the criteria for acute pancreatitis with serum lipase elevated at greater than three times the upper limit, alongside epigastric pain radiating to the back.

Treatment is similar to other presentations of acute pancreatitis, with IV fluids, pain control and electrolyte repletion. The clinical significance of early diagnosis relies on the indication for exchange blood transfusion in worsening cases of acute pancreatitis characterized by multi-organ failure\(^6,8,9\) which helps dilution of Hemoglobin S and reduction in sickling with hemoglobin S goal less than 20-30%\(^10\).

References

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The Heroin Epidemic Forbodes the Acquired Gerbode Defect

Authors: Vybhav Jetty MD, Steven Rudick MD, David M. Harris MD

Introduction: Gerbode defect is a rare anomaly that connects the left ventricle (LV) to the right atrium (RA). As a congenital lesion, the connection was first reported in 1838. While first discovered as a congenital defect, the entity can also be acquired. As an acquired defect, the Gerbode defect is increasingly prevalent as a complication of endocarditis, myocardial infarction, or previous cardiac surgery.

Case Presentation: The patient is a 34 year old female with history of Enterococcal endocarditis and heart block with pacemaker dependence. One year after her initial endocarditis, she presented with aortic valve endocarditis due to Serratia, aortic root abscess, severe aortic insufficiency, and right frontal embolic stroke. Intraoperatively, her root abscess was unroofed and debrided. Her left ventricular outflow tract (LVOT) was closed using a pericardial patch. The valve was replaced with a bioprosthetic pericardial valve. Several days later, she returned with new onset congestive heart failure, right pleural effusions, and change in her murmur. Transthoracic echocardiography revealed an LVOT pseudoaneurysm and fistula connecting the LV to the RA. Transesophageal echocardiography confirmed the mitral-aortic intervalvular fibrosa pseudoaneurysm. This pseudoaneurysm and fistulous tract extended from the LVOT, involving the aortic root and surrounding structures, eventually creating and left-to-right shunt into the RA. The defect was repaired pericardial patch.

Discussion: Diagnosis is made by transthoracic and transesophageal echocardiography. The pressure gradient between the LV and RA results in high velocity Doppler flow signal in the RA and should not be mistaken for tricuspid regurgitation. Acquired LV-RA shunts are more often associated with multiple comorbidities, including congestive heart failure, valvular leaflet perforation, subannular abscess, and complete heart block. Surgery for the shunt is important as these subtypes cannot be fixed percutaneously in the setting of infection. Recent reports describe an increase in the total number of cases of acquired Gerbode defects due to endocarditis. Give the recent rise in endocarditis from the heroin epidemic, the incidence of Gerbode defect will likely continue to rise. Currently there are no recommendations for this defect, however surgical closure is the treatment of choice. However, percutaneous closure has been described in the literature with good outcomes in high-risk candidates. Asymptomatic, chronic, small defects can be managed conservatively.
Introduction: Household bleach is used worldwide as a cleaning and disinfecting agent. Excluding caustic injury from oral ingestion, common untoward effects from household bleach exposure include cough, irritation of eyes, nose, and throat, and nausea. Reactive airways dysfunction syndrome has also been reported.1 In general, untoward effects are self-limited. We report a case of acute respiratory distress syndrome (ARDS), and eventual death, related to household bleach exposure. This case emphasizes the importance of public health awareness and utilization of in-home protection strategies when using bleach-containing cleaning products.

Case Presentation: A 75-year-old non-smoking female with no cardiopulmonary disease presented to an outside hospital with dyspnea. She lay next to a spilled gallon of bleach for approximately 2 hours following a mechanical fall while cleaning her bathroom. On-scene medics noted oxygen saturation of 70% on room air. She did not improve with BiPAP, required elective intubation, and was transferred to our facility. Chest CT showed diffuse infiltrates consistent with ARDS. Symptom severity prompted bronchoscopy which was unremarkable and she was diagnosed with acute hypoxemic respiratory failure secondary to moderate ARDS from caustic bleach inhalation. Initial PaO2 / FiO2 ratio was 61 and minimally improved to 100-150 with steroids, diuresis, empiric antibiotics (despite no identifiable superimposed infection), and ventilatory support. She failed attempts at weaning from ventilator support, compassionate extubation was elected, and she expired on Day 8.

Discussion: Severe respiratory toxicity from chlorine gas exposure is usually reported in association with significant industrial incident or swimming pool spills.1 Acute inhalation and chronic exposure to household bleach have been associated with bronchial hyper-responsiveness, asthma and chronic bronchitis,2 however severe pulmonary toxicity is rare.3,4 Few cases of life-threatening toxic pneumonitis, ARDS and respiratory failure are reported.3,4,5,6 Extent and severity of lung injury depends on duration of exposure and minute ventilation of the person exposed.4 Respiratory insult can occur with mixing of bleach and ammonia and formation of chloramine compounds, from formation of hypochlorous acid and reactive oxygen species,7 and release of free radicals from neutrophils recruited to airways after acute chlorine inhalation.4 Management is supportive, including source removal, oxygen supplementation, and bronchodilator administration.4 Superimposed pulmonary infection occurs in approximately 50% of affected individuals;8 though our patient received empiric antibiotics, no infection was documented. The role of inhaled or systemic steroids is controversial given the risk of superimposed infection, however animal studies and few case reports suggest improved mortality rates with steroid use in acute chlorine exposure.4

References

Connecting the dots – Acute abdomen, ST elevation and a consolidating lung mass

Authors: Randol Kennedy MD, Percy Adonteng-Boateng MD, MPH

Introduction: Community acquired pneumonia is a commonly treated disease and yet may present in very unfamiliar ways that if missed may result in unnecessary testing, procedures and potential harm. Encouraging reporting and discussion of these atypical presentations may help increase clinical suspicion and timely diagnosis.

Case Presentation: We present a 38 year old male with no prior medical history who came to the ED with a three day history of severe, cramping lower abdominal pain. This was intermittent, non-radiating, and was exacerbated with movement. Associating symptoms were anorexia, constipation, fever, chills, nausea and retching. He denied use of tobacco, alcohol or recreational drugs.

Physical exam revealed an acutely ill patient who was febrile (38.8°C), with pulse 89 beats/min, respiratory rate 18 cycles/min, BP 164/85 with orthostatic changes and oxygen saturation (room air) 98%. On abdominal exam, there was generalized abdominal tenderness with rigidity and guarding. Respiratory exam was benign. The patient was therefore worked up for an intra-abdominal cause of acute abdomen which included acute appendicitis. Labs drawn showed white blood cell count 11300/microL without bands and hemoglobin 12.9 g/dl. CT scan of the abdomen revealed normal abdominal viscera. However, a “mass like opacity” in the right lower lobe of the right lung was reported. Meanwhile, a 12 lead EKG showed left ventricular hypertrophy, with significant ST elevation in the anterior leads, pathological q waves, T wave inversion in the lateral chest leads as well as reciprocal changes in the inferior leads. Cycled troponins were negative.

Review of systems by the admitting team highlighted a history of intermittent productive cough with yellow sputum one day prior to presentation. Dullness to percussion over right posterior lower lung fields, was also noted on exam. A CT scan of the lung confirmed right lower lobe consolidation. Pneumonia hence became the diagnosis, with an atypical presentation of acute abdomen. The patient was admitted and treated with ceftriaxone and azithromycin. On day two, lung crepitations became evident, which resolved prior to discharge. Serological and microbiological tests for legionella, mycoplasma, influenza and blood cultures were negative. Transthoracic echocardiogram was unremarkable.

Discussion: Abdominal pain as a presenting feature of community acquired pneumonia is predicted to be found in 8% of cases, with the full picture of pneumonia occurring later in the diagnosis. EKG abnormalities - also an uncommon feature in - is independent on the organism and can mimic any alarming pathology. Awareness of the chameleon potential of common diseases such as community acquired pneumonia can sharpen our clinical acumen, reminding us the importance of the utilization of a thorough review of systems as well as a contextual approach to diagnosis.

References

Elevated International Normalized Ratio with Apixaban in Patient with End-Stage Renal Disease on Hemodialysis

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Introduction: Apixaban can prolong International Normalized Ratio (INR). Changes in INR are dose related and occur about 3 hours post dosing. While apixaban dosage labeling includes patients with atrial fibrillation and severe renal impairment, there are no recommendations for any other indication for patients with chronic kidney disease (CKD) stage V on hemodialysis.

Case Presentation: A 68-year-old Hispanic male (60.2 kg) was referred to the emergency department (ED) for elevated PT/INR that was done as part of admission testing panel to a rehabilitation facility for ongoing management of surgical repair and internal fixation of a bimalleolar right ankle fracture. Past medical history included CKD stage V on hemodialysis, cirrhosis and chronic hepatitis C.

His medication review included apixaban 2.5 mg twice daily for 3 days (6 doses) for venous thromboembolic event prophylaxis. One day prior to surgery INR was 1.2. At the rehabilitation facility an INR of 2.74 was noted and in the ED was recorded as greater >18 with protime of >200 seconds. Patient denied any genitourinary or gastrointestinal bleeding. No acute neurological symptoms were reported.

On physical examination, he had a blood pressure of 151/57 mmHg, a grade III/IV systolic murmur radiating to the axilla and an ecchymotic discolorations at the surgical site. As per ED protocols, patient was given 10 mg of Vitamin K intravenously and with decrease of INR to 2.0 five hours later. Mixing studies revealed a PT that did not fully correct suggesting possible invivo or exvivo circulating inhibitors. On the subsequent day the INR has normalized to 1.1. Patient was discharged back to the rehabilitation facility on 1mg of Warfarin daily.

Discussion: Significantly elevated INR of >20 with apixaban is rare as opposed to rivaroxaban. Vitamin K is not expected to neutralize the effect of apixaban and its administration may not have had any beneficial effect on this case. In a patient dosed with apixaban, INR is not indicated for monitoring of drug efficacy or toxicity. Therefore, in the absence of any major bleeding, careful monitoring may be all that is required. Andexanet alfa has been approved for reversal of anticoagulant effect of apixaban in cases with uncontrolled or life-threatening bleeding. Furthermore, there is very little data to support the uses of apixaban in patients with CKD stage V on hemodialysis for venous thromboembolic thromboprophylaxis involving lower leg injuries requiring leg immobilization. Further studies are needed and are necessary to further elucidate the clinical significance such INR values associated with apixaban and other direct oral anticoagulants.

References


It’s Just a Cold Sore: Acute Liver Failure Secondary to Disseminated Herpes Simplex Virus due to Ixekizumab

Authors: Mark Saling, DO. Muhammad Ahsan Zafar, MBBS.

Introduction: Acute liver failure (ALF) is typically caused by hypotension, toxins or viral hepatitis. Viral hepatitis is typically caused by hepatitis virus A through E and rarely caused by viruses such as Cytomegalovirus, Epstein-Barr virus and herpes simplex virus (HSV). HSV 1 typically presents with oral ulcers, while HSV 2 presents as genital ulcers. HSV Hepatitis is a rare cause of ALF that carries a staggeringly high mortality rate due to poor recognition.

Case Presentation: 47-year-old male presented with altered mental status, septic shock and significantly elevated transaminases. The patient had a history of psoriasis and had recently been tapered off ixekizumab due to GI upset and started on systemic prednisone. The patient had negative toxicology screen, negative blood, urine and cultures and unremarkable CSF studies. He was started on drug spectrum antibiotic and antifungal coverage, but the patient significantly declined requiring intubation, continuous renal replacement therapy, and multiple vasopressors. Although no active lesions were discovered, the patient was found to have positive serum HSV 1. Despite initiation of acyclovir and aggressive liver transplant evaluation, the patient continued to decline and care was withdrawn.

Discussion: Approximately 80% of the US population has HSV 1 with outbreaks increasing during periods of high stress and immunosuppression. Despite near pandemic HSV, disseminated HSV infections are a rare cause of acute liver failure typically occurring in immunocompromised states as well as during pregnancy. Early recognition of HSV induced ALF is critical given an 80% mortality rate if left untreated and 50% mortality rate even with treatment with acyclovir. However, HSV is not included in the initial screening test. A high index of suspicion should be maintained in all patients presenting in acute liver failure who are immunocompromised or culture negative and have negative toxicology screens. With an increase in the frequency of HSV hepatitis in the advent of new immunotherapy biologic agents, the diagnostic workup pathway must evolve to include HSV if we hope to recognize HSV hepatitis in time to successfully treat it.
Rapid Progression of Ischemic Cardiomyopathy: Beyond the Pale Tissue

Authors: Alexander Smith, DO; Kim Jordan MD, Department of Internal Medicine - Riverside Methodist Hospital, Columbus, OH

Introduction: Cardiac sarcoidosis is well-described, but often remains undiagnosed. Cardiac involvement is only clinically apparent in approximately 5% of sarcoidosis patients, but one study reported autopsy detection in 25% of patients with known sarcoid disease.

Case Presentation: A 49-year-old male with known ischemic heart disease and sarcoidosis was admitted for repeated ICD firing due to ventricular fibrillation storm. He presented 11 months prior following cardiac arrest with ventricular fibrillation, requiring multiple defibrillation attempts to restore circulation. Evaluation showed ST-elevation myocardial infarction (MI) and echocardiography showed ejection fraction (EF) 40-45% with multiple wall motion abnormalities. Cardiac catheterization revealed 100% mid-LAD thrombosis, (no significant disease in other coronary vessels), and a drug-eluting stent was placed. Over the next 6 months, his EF declined to 35-40%, and an implantable cardiac defibrillator was placed. He started cardiac rehabilitation, but was subsequently admitted for acute heart failure, believed secondary to dietary non-compliance. Repeat echocardiography, however, showed EF 25-30%. Shortly thereafter, he developed aberrant ICD firing, new-onset atrial fibrillation, and required anti-arrhythmic therapy. However, aberrant ICD firing persisted requiring 2 additional hospitalizations and atrial lead placement for overdrive pacing. His EF was now 15-20%. On this admission, heart failure specialists questioned whether disease progression and arrhythmia were related to ischemic disease. Cardiac MRI showed extensive abnormal myocardial enhancement in a non-ischemic pattern in the basal, mid-anterior, anterolateral, inferolateral, basal anteroseptal and right ventricular walls, consistent with cardiac sarcoidosis. Corticosteroid and immunomodulating medications were initiated. Along with additional antiarrhythmics, he has not had further ventricular arrhythmias.

Discussion: An estimated 16-35% of patients with complete AV block (<age 60) or ventricular tachycardia of unclear origin have undiagnosed cardiac sarcoidosis. Manifestations of cardiac sarcoidosis include arrhythmias, ventricular hypertrophy, intraventricular septal thinning, diastolic and systolic dysfunction, and wall motion abnormalities. Symptom severity is variable and symptom absence does not exclude disease. International consensus guidelines state that cardiac sarcoidosis is diagnosed by the presence of non-caseating granulomas in myocardial tissue. Probable diagnosis is established with histological diagnosis of extracardiac sarcoidosis, exclusion of other causes, and one or more of the following: steroid and immunosuppressant responsive cardiomyopathy or heart block, unexplained LVEF<40%, unexplained sustained ventricular tachyarrhythmia, a Mobitz type II second or third-degree heart block, patchy uptake on dedicated cardiac PET imagine, late gadolinium enhancement on cardiac MRI (in pattern consistent with cardiac sarcoidosis). Our patient had biopsy-proven extracardiac sarcoidosis many years prior to presentation. While we cannot exclude myocardial scarring from his infarction as the cause of his arrhythmias, his cardiac MRI findings and history are consistent with probable cardiac sarcoidosis. This case illustrates that providers should “think outside the box” when evaluating progressive cardiac disease in a patient with both ischemic heart disease and sarcoid disease.

References

Eiploic appendagitis: An often misdiagnosed cause of acute abdomen

**Authors:** Vishnu Charan Suresh Kumar MD, Hisham Alwakka MD, James Shina MD

**Introduction:** Epiploic appendagitis is an uncommon clinical entity that causes acute abdominal pain. It is often misdiagnosed as acute diverticulitis or appendicitis. Here, we report a case of a 72-year-old lady who presented with acute left lower quadrant abdominal pain. The purpose of this case report is to raise awareness of such a clinical condition with its characteristic imaging findings. This is very important to avoid hospitalization costs, additional antibiotic course, and the morbidity and mortality associated with surgical procedures.

**Case Presentation:** A 72-year-old lady with a past medical history of SVT, hypertension, and dyslipidemia presented to the ER complaining of localized left lower abdominal pain for two days, sudden onset, constant, progressive, 8/10 in intensity and without any relieving or aggravating factors. She denied fevers, chills, nausea, vomiting, anorexia, urinary symptoms or change in bowel habits. She has never had appendicitis or diverticulitis.

On examination, the patient was comfortable, not in distress. She was afebrile, vital signs were within normal limits, and abdominal examination was positive for localized tenderness on palpation in the left lower quadrant. Laboratory studies were unremarkable. A contrast-enhanced CT of the abdomen and pelvis was obtained and reported to show acute uncomplicated diverticulitis. The patient was admitted and started on IV antibiotics and analgesics.

Upon review of CT scan during rounds on day 2, there was a fat-density ovoid lesion with hyper-attenuation center abuts the anterior wall of the proximal sigmoid colon measuring about 20x10x10 mm, mild thickening of the adjacent colonic wall and mild stranding of the adjacent fat. The constellation of these classic findings on CT is considered characteristic for acute epiploic appendagitis(Image1.1). IV antibiotics were discontinued, and the patient did well and had complete resolution of her symptoms within 48 hours of her initial presentation. The patient was given reassurance and discharged home on NSAIDs. She followed up with us in a week and was doing well.

**Discussion:** Epiploic appendagitis is a rare benign clinical entity that causes acute abdominal pain that was first described in 1919 by Hunt. Retrospective studies have found its incidence to be 8.8 per million people. Epiploic appendages are fat-filled, serosa-covered structures positioned in two separate longitudinal lines along the colon. Torsion or thrombosis of the appendageal draining vein causes inflammation and peritoneal irritation which leads to localized abdominal tenderness and even guarding in some cases. Given the lack of typical symptoms and signs, it is often misdiagnosed as either appendicitis or diverticulitis depending on the location and treated accordingly. Awareness about this entity amongst clinicians and identification of typical CT findings by radiologists upon initial presentation would help reduce unnecessary antibiotic use, diagnostic testing and in some instances unwarranted surgery.
Safe Use of Rituximab in Pregnancy for Polymyositis: A Case Report

Authors: Timothy Underwood, DO; Matthew Mundwiler, MD; Kim Jordan, MD

Introduction: Controlling autoimmune disease activity during pregnancy is associated with less peripartum and neonatal complications1. Polymyositis is an uncommon inflammatory myopathy affecting predominantly females during child-bearing years. Antisynthetase syndrome, a subset of polymyositis, is characterized by polymyositis, interstitial lung disease (ILD), mechanic’s hands, and Raynaud’s phenomenon. For patients who are pregnant and have uncontrolled disease after first-line treatments, choosing a second-line agent poses a challenge, as most agents are known teratogens2. Rituximab has been shown to be effective for polymyositis, however, to date, the safety of rituximab during pregnancy is unclear. Here we present a case of rituximab use during pregnancy to maintain remission in refractory polymyositis.

Case Presentation: A 31-year-old female with antisynthetase syndrome presented to the rheumatology clinic after learning she was pregnant. Rheumatologic disease was diagnosed one year prior when she developed recurrent episodes of bibasilar pneumonia and generalized weakness. Extensive evaluation was significant for CRP 61.5 mg/L, ESR 59 mm/hr, total CK 684 U/L, aldolase 21.2 U/L, positive SSA >100, and positive PL12. Pulmonary function tests showed reduced diffusion lung capacity and mild restriction, but bronchoscopy was non-revealing. Lung biopsy was interpreted as organizing pneumonia. Myopathy work-up included electromyography which was negative, and magnetic resonance imaging of the femurs that was positive for bilateral edema in vastus lateralis muscles. Thigh muscle biopsy was consistent with polymyositis. Antisynthetase syndrome was diagnosed given the constellation of findings.

The patient was treated with high-dose steroids and improved, though failed steroid taper and trial of azathioprine. Taking into account plans for future pregnancy, she was initiated on rituximab with good response. Six months after infusion, the patient was found to be 15 weeks pregnant. Following a literature review, risks and benefits of continued rituximab therapy were discussed with the patient. Given the extent of her illness, she elected to continue rituximab, which was re-dosed at week 19. She had no disease flares and delivered a healthy baby at 39 weeks 2 days gestation with no obstetric or neonatal complications.

Discussion: Experience with rituximab during pregnancy has been documented in case reports. Chakravarty et al. described 20 cases of drug administered during pregnancy for lymphoma or life-threatening autoimmune blood disorders3. Neonatal adverse events included 6 with hematologic abnormalities (neutropenia, lymphopenia, B cell depletion) and 2 others with infection (CMV hepatitis transmitted from mom, bronchiolitis). Ostensen et al. described 4 similar cases that resulted in all neonates with lymphopenia or B cell depletion, though no associated complications4. Unlike the above documented use in life-threatening disease, this case shows successful use of rituximab during pregnancy for remission in polymyositis. With more data on safety and use, rituximab may begin to play a key role in disease remission during pregnancy for rheumatic disease.

References

OKLAHOMA CLINICAL VIGNETTE POSTER FINALIST - BLAKE A JACOBS, MD

HYDRALAZINE-INDUCED ANCA VASCULITIS IN A PATIENT WITH UNDERLYING CHRONIC KIDNEY DISEASE: REPORT OF A DIAGNOSTIC CHALLENGE

Authors: Blake Jacobs MD, Satish Kumar MD

Introduction: Hydralazine is direct-acting arterial and arteriolar vasodilator commonly used in the treatment of hypertension and heart failure. While hydralazine-induced lupus is more commonly reported, there is an increasing body of literature regarding the rare complication of hydralazine-induced antineutrophil cytoplasmic antibody (ANCA) vasculitis. Diagnosis of hydralazine-induced ANCA vasculitis is often complicated by the rarity of the diagnosis, overlap with primary small-vessel vasculitides, and non-specificity of symptoms.

Case Presentation: A 70-year-old African American gentleman with a greater than 10-year history of chronic kidney disease (CKD) stage 3 from poorly controlled diabetes mellitus and hypertension presented to our facility for kidney biopsy. He had been discharged several days prior following a 10-day admission for hemoptysis and cough. Chest x-ray at that time showed right sided infiltrates concerning for community acquired pneumonia treated empirically with levofloxacin with resolution of symptoms. He was noted on that admission to have an acute kidney injury (AKI) with an increase in creatinine from baseline of ~2.8 mg/dL to 3.6 mg/dL initially felt to represent dehydration. Despite improvement of pulmonary symptoms, kidney function continued to decline. Workup included a normal renal ultrasound, normal kappa/lambda ratio, negative anti-nuclear antibodies, negative HIV and hepatitis panels, normal complement levels, and negative anti-glomerular basement membrane antibodies. He was discharged but returned for outpatient biopsy. At that time previously pending labs were reviewed that included an anti-MPO-ANCA by enzyme-linked immunoassay strongly positive at >100 units/mL [ref: 0-9], perinuclear ANCA titre of 1:160 [ref: <1:20], and anti-histone antibody strongly positive at 6.4 units [ref: 0.0 – 0.9]. A review of outpatient medications at that time revealed that the patient had been receiving oral hydralazine for blood pressure control for 2 years with a recent dose increase from 50mg three times a day to 100mg three times a day one month before the onset of hemoptysis and worsening renal function. The pathology from the renal biopsy later confirmed necrotizing pauci-immune glomerulonephritis consistent with hydralazine-induced ANCA vasculitis.

Discussion: Although rare, hydralazine causing ANCA-vasculitis with resultant renal and pulmonary complications is well-documented. As our case shows, without a high index of suspicion, the symptoms of drug-induced vasculitis can be easily overlooked. Recognition of this possible devastating reaction to hydralazine can lead to prompt diagnosis and intervention to avoid long-term harm to the patient.

References

**OKLAHOMA CLINICAL VIGNETTE POSTER FINALIST - JANITZIO J GUZMAN, MD**

**Pasteurella multocida peritonitis in a patient on peritoneal dialysis: a reminder on personal hygiene and pets**

**Authors:** Janitzio Guzmán, MD; Rui Mao, MD; Krishna Baradhi, MD

**Introduction:** End-stage renal disease (ESRD) affects more than a million Americans. While the majority of patients receive hemodialysis, less than 10% receive peritoneal dialysis (PD). Though the overall rate of peritonitis among these patients is 0.65 episodes/patient-year, it continues to be the major complication, cause of morbidity, and a barrier to PD. *Pasteurella multocida* infection remains a rare but significant cause of peritonitis. We report a case of *Pasteurella multocida* peritonitis in a patient on PD and emphasize the importance of personal hygiene to prevent zoonotic infections.

**Case Presentation:** A 56-year-old man with Hepatitis C and ESRD on cycler PD presented with abdominal pain, vomiting and cloudy peritoneal fluid. Physical exam was remarkable only for mild abdominal tenderness. CT abdomen was unrevealing. Initial gram stain of peritoneal fluid was negative but showed turbid fluid with WBC count of 7175/mm³ with 90% polymorphonuclear cells. The culture later grew *Pasteurella multocida*. Retrospective history and home inspection revealed close contact with a domestic cat and poor hygiene as a potential source of this zoonotic infection. He was re-educated about home PD care and was treated with intraperitoneal ceftriaxone, resulting in steady resolution of his symptoms.

**Discussion:** PD peritonitis is usually a monomicrobial infection, secondary to Gram-positive cocci or enteric Gram-negative bacilli. This case of peritonitis due to *Pasteurella multocida* is a rare etiology of a common problem among PD patients and demonstrates that these high-risk patients may be susceptible to unusual zoonoses. Our case highlights the importance of patient education about transmission of infection from pets as well as a thorough evaluation of risk factors for exposure. Championing patient education through a multidisciplinary approach is crucial for favorable outcomes and to prevention of zoonotic infections.

**References**

A RARE CASE OF DIFFUSE ALVEOLAR HEMORRHAGE ASSOCIATED WITH ANTI-SYNTHETASE SYNDROME LEADING TO ACUTE HYPOXIC RESPIRATORY FAILURE

Authors: Anam Siddiqui, MD, Fatima Sukhera, MD, Juan Antonio Cantu, MD, Matlock Jeferies, MD

Introduction: Anti-synthetase syndrome is a clinical entity characterized by specific anti-aminocyl-tRNA-synthetase antibodies usually associated with inflammatory myopathy, interstitial lung disease, characteristic skin findings, Raynaud’s phenomenon, and arthritis. The classic presentation of the pathology is rapidly-progressive interstitial lung disease, which commonly determines the prognosis. The diagnosis of anti-synthetase syndrome in patients with acute respiratory distress syndrome (ARDS) is unusual, however not uncommon.

Case Presentation: Here we present a case of a 44 year-old Hispanic male with recent diagnosis of BAL proven diffuse alveolar hemorrhage who presented to a tertiary care hospital with dyspnea, pleuritic chest pain, room air hypoxia, blood tinged hemoptysis, lower extremity swelling, fevers/chills, night sweats and a 20lbs weight loss. He was started on 12L oxygen via oxymizer at the time of admission and continued to have worsening respiratory distress thus requiring intubation due to acute hypoxic respiratory failure. Subsequent chest radiography showed steadily worsening pulmonary disease characterized by white-out of bilateral lung fields, and CT chest demonstrated multi-lobar ground glass opacities. Given our concern for both infection and autoimmune disease, the patient was started on broad-spectrum antibiotics, pulse steroids and 5 days of plasmapheresis. Pulmonary Tuberculosis was ruled out. Repeat bronchoscopy was performed to rule out recurrent DAH (none found, but diffuse GGO consistent with ARDS persisted on high res chest CT). An antinuclear antibody test by indirect immunofluorescence demonstrated a high-titer cytoplasmic and nucleolar pattern (1:9720 titer), and specific autoantibody testing revealed both anti-Ro and anti-PL-7 antibody specificities. Given this, a diagnosis of antisynthetase syndrome-related ILD with the unusual initial presenting feature of DAH was made. B-cell depletion therapy with 1g Rituximab was given and an oral corticosteroid taper was initiated, with rapid improvement in his clinical symptoms. He was discharged home on 3 L oxygen via NC with a steroid taper and plans for close follow up with Rheumatology for Rituximab infusion and Pulmonology for management of ILD.

Discussion: Early diagnosis of anti-synthetase syndrome followed by appropriate treatment can help prevent serious long term complications in patients. It is thus critical for physicians to keep relatively unusual causes of autoimmune lung disease in the differential diagnosis of such patients, and to perform early testing in patients with high clinical suspicion of disease.

References

Stop Calling Me Crazy! – The Unusual Course of NMDA-Receptor Encephalitis

Authors: Vassilyadi, P, MD; Shiari, A, MD; Hughes C, MD, FACP

Introduction: Over the past decade there have been increasing reports of autoimmune encephalitis, most commonly caused by anti-NMDA-receptor antibodies. It is predominantly described in women and can be associated with ovarian teratoma. We present a case of anti-NMDA-receptor encephalitis from onset to complete resolution.

Case Presentation: A 54-year-old female with no medical history presented with status epilepticus. Two weeks earlier she had been admitted to a psychiatric unit for acute psychosis-like symptoms and personality change. Her mentation deteriorated until she became catatonic, developed seizures and was intubated due to unresponsiveness. Lumbar puncture was performed and cerebrospinal fluid (CSF) demonstrated lymphocytic pleocytosis, oligoclonal bands and anti-NMDA-receptor antibodies. Brain MRI and whole-body CT were normal. EEG showed severe encephalitis without epileptogenic foci. She received five days of high-dose solumedrol and five days of intravenous immunoglobulin. She remained encephalopathic with oral-lingual-facial dyskinesia and dysautonomia. She began treatment with weekly Rituximab, which improved mentation. After four weeks of therapy, repeat CSF studies demonstrated absence of anti-NMDA-receptor antibodies. After several weeks of rehabilitation, symptoms completely resolved and she returned to normal activities of daily living.

Discussion: The complete clinical course of NMDA-receptor encephalitis is not well documented. Clinical presentation varies from acute psychosis to status epilepticus, coma and death. This disorder can easily be confused with acute psychosis and many patients are placed in psychiatric facilities. It is imperative for the clinician to recognize this condition as non-psychiatric and initiate prompt therapy with intravenous steroids and immunoglobulin. Severe cases have responded to Rituximab.
Elsberg syndrome: a rare infectious cause of cauda equina syndrome

Authors: Joel Burnett, MD; Christopher Terndrup, MD

Introduction: Cauda equina syndrome (CES) is an important neurologic disorder characterized by incontinence, paresthesias, and sexual dysfunction. Etiologies of CES include intervertebral disk herniation, chronic lumbosacral stenosis, trauma, tumors, and viral infections. We present a rare case of CES due to a viral infection.

Case Presentation: A 62-year-old female with chronic left lower extremity radiculopathy, chronic kidney disease, hypertension, and remote invasive ductal carcinoma in remission presented to urgent care with one week of chills, vaginal pain, dysuria, urinary retention, and constipation. She had a new sexual partner of 4 months. They did not use condoms. On exam, vitals were within normal limits. Genitourinary exam revealed vaginal erythema and multiple, bilateral ulcerative lesions of the labia. Urinalysis was unremarkable. Tests for *Neisseria gonorrhoeae* and *Chlamydia trachomatis* were negative. HSV culture of a genital lesion was positive. She was prescribed acyclovir and advised to follow up in one week. A week later she represented with improvement in the genital lesions and new complaints of perineal numbness and fecal and urinary incontinence. A detailed neurologic exam was not documented. CT abdomen and pelvis with contrast revealed anterolisthesis of L3-L4 causing narrowing of the thecal sac. After 6 weeks, she presented to her primary care physician (PCP), reporting persistent but improved fecal incontinence and perineal paresthesia. Exam was notable for normal gait, 5/5 lower extremity strength, decreased perineal sensation, and flaccid rectal tone. She was sent to the emergency department for urgent MRI and surgical evaluation. MRI spine without contrast redemonstrated L3-L4 anterolisthesis with moderate canal stenosis. She was evaluated by orthopedic surgeons who concluded that “her degree of stenosis [did] not explain her incontinence symptoms. Other causes should be investigated” thus she was discharged. After 3 weeks, she saw her PCP and reported continued improvement in fecal incontinence and paresthesia. After careful review of primary data and consideration of etiologies of CES, she was diagnosed with Elsberg syndrome – a self-limited cause of CES due to genital HSV infection – and was continued on acyclovir.

Discussion: In this case, a woman with chronic spinal stenosis developed subacute CES after acute HSV infection, underwent spinal imaging and surgical evaluation, and was judged to have insufficient spinal stenosis to account for CES. Over time she experienced spontaneous symptom improvement. This presentation is best explained by Elsberg syndrome, a manifestation of a primary infection or reactivation of HSV-2 consisting of acute/subacute sacral radiculitis. The syndrome presents with CES of acute/subacute onset and recent or coexisting HSV infection in the absence of alternative CES causes. The syndrome is likely under-recognized. Epidemiological data is scarce. While managing patients with acute or chronic genital HSV, physicians should be aware of this severe aspect of the clinical spectrum of genital herpes infection.
Double Trouble in A Bariatric Patient

Authors: Ashray Maniar MD, Deanna Green, MD, Alan J. Hunter, MD, FACP

Introduction: As bariatric procedures become more common, the prevalence of post-procedure complications increases, including both mechanical and nutritional complications. This case highlights a severe thiamine deficiency that developed after prolonged reflux and emesis after a sleeve gastrectomy.

Case Presentation: A 23-year-old Saudi Arabian woman with a history of obesity and 10 weeks status post sleeve gastrectomy, presented with acute onset diplopia, lightheadedness, and gait imbalance in setting of persistent, intractable post-operative nausea and vomiting. Initial vital signs and electrocardiogram were normal. Physical exam was notable for bidirectional, coarse horizontal nystagmus, diplopia in multiple visual fields, atactic gait, and normal mental status. Initial studies revealed a negative urine pregnancy test, potassium of 2.7 mmoL/L (normal 3.4-5.0), BUN 2 mg/dL (6-20), creatinine 0.34 mg/dL (0.6-1.1), and albumin 3.2 g/dL (3.5-4.7). A head CT was normal and CT abdomen showed chronic fluid collection adjacent to the gastric remnant. A cranial MRI was unremarkable. Given the patient’s diplopia, nystagmus, and ataxia, empiric high dose intravenous thiamine (500 mg TID) was initiated for presumed acute post-bariatric surgery Wernicke encephalopathy. We considered if the fluid filled gastric remnant was contributing to her nausea, however, with its small size and chronicity, this was deemed unlikely. Further lab testing revealed multiple water and fat-soluble vitamin deficiencies. Her visual deficits and gait disturbances rapidly resolved with thiamine repletion. She was educated on the importance of vitamin supplementation and a bariatric diet and safely discharged.

Discussion: The increasing prevalence of obesity in the United States parallels the rise in performance of bariatric surgeries. Thus, it is important that clinicians recognize and manage both immediate and latent post-surgical complications. Common complications following both malabsorptive and restrictive bariatric surgery include reflux, dumping syndrome, and micronutrient deficiencies that can manifest early or late following surgery. Without frequent education, appropriate supplement regimens, and patient/physician vigilance these deficiencies can be severe. Thiamine, a key cofactor in multiple cerebral metabolic pathways, is one such micronutrient that becomes deficient in up to 30-49% of bariatric patients within days-months after surgery. Deficiency causes neuronal disruption and, potentially, necrosis—causing a classic triad of altered mental status, vision changes (ophthalmoplegia, diplopia, nystagmus) and gait disturbances as well as temperature dysregulation and hypotension. The diagnosis is clinical and based on risk factors with any of the above clinical signs, as the triad is present in only 10-16% of cases. Untreated, it can lead to permanent neurologic damage and death, mandating urgent treatment. Treatment should include parenteral thiamine, to ensure adequate delivery across the blood-brain barrier, with recommendations varying widely in dose (50 mg-1 G) and frequency (qD-TID) due to a paucity of data. However, there is exceedingly low evidence of toxicity at high doses.

References

This case underscores the importance of recognizing and managing post bariatric surgery nutritional deficiencies and specifically the clinical syndrome of thiamine deficiency, previously described as bariatric beriberi.
Usual Case of Acute Alcohol Withdrawal Leading to an Unusual Case of Cardiomyopathy

Authors: Navneet Kaur MD, Sowjanya Yenigalla MD, Fnu Abhishek MD, Kaur MD.

Introduction: Takotsubo Cardiomyopathy (TCM), also known as stress cardiomyopathy or apical balloons syndrome, is a transient decrease in left ventricular function, reported in 1-2% of patients presenting with symptoms similar to acute coronary syndrome. The trigger for TCM is thought to be an acute emotional or physical stressor causing a surge in catecholamine release. Acute alcohol withdrawal state has been linked to TCM in some cases.

Case Presentation: 65-year-old male with history of hypertension, diabetes mellitus, alcohol abuse and paroxysmal atrial fibrillation presented with complaints of epigastric pain, nausea, emesis and decrease in appetite for five days. He has a history of drinking one pint of alcohol daily, last drink being two days prior to admission. On initial presentation, an electrocardiogram (ECG) demonstrated sinus tachycardia with ventricular rate of 112. Overnight, he developed atrial fibrillation with rapid ventricular rates, confirmed by ECG. On admission, an alcohol withdrawal protocol was initiated. Initial troponin enzyme levels were negative. An echocardiogram showed normal left and right ventricular size and function. Ejection fraction of 60-65%. On day five, he developed retrosternal chest pressure that was relieved by nitroglycerin. Vitals signs obtained while symptomatic were significant for hypertension to 175/105mmHg, normal heart rate, however, his extended release beta-blocker dosage was doubled thirty-six hours prior to this event. ECG showed sinus rhythm, deep symmetrical T wave inversions in anterior septal and apical leads. Troponin level elevated, peaked at 0.11 ng/ml. Based on chest pain, dynamic ECG changes and mildly elevated troponin enzymes, cardiac catheterization was warranted. It demonstrated moderate non-obstructive disease of coronary arteries. Left ventriculogram showed apical near akinesis and preserved basal wall motion, characteristic of TCM. Ejection fraction 35-40%. Throughout admission, patient was receiving multiple doses of benzodiazepines for elevated withdrawal assessment scores. Patient was discharged home with Cardiology out-patient appointment.

Discussion: TCM can be an unusual complication of an acute alcohol withdrawal state. Patients complaining of chest pain with signs and symptoms of alcohol withdrawal should be carefully assessed as there is a risk of catecholamine surge and sympathetic hyperactivity, which may lead to a pathophysiologic phenomenon similar to that of TCM.

References

Introduction: Acute kidney injury is a common complication seen in hospitalized patients. It has been associated with morbidity, mortality, increased length of stay and healthcare cost. We describe a patient with a complicated hospital course who suffered an acute kidney injury of uncommon etiology and remains dialysis dependent.

Case Presentation: A 78 year old male with a history of CAD, HTN and DMII presented for CABG surgery. Post-operatively, he developed sustained MRSA bacteremia secondary to sternal wound infection and was treated with vancomycin. He was also found to have an upper extremity superficial venous thrombosis and was started on warfarin. During his stay, he suffered an acute kidney injury with serum creatinine of 2.0mg/dl from baseline 0.8mg/dl. It was thought to be secondary to vancomycin toxicity and he was transitioned to daptomycin. Despite cessation of vancomycin, he continued to have worsening renal function with significant proteinuria; up to 20g/day. An extensive work up including SPEP, UPEP, complement levels, hepatitis B and C serologies, HIV, ANCA, and ANA was unrevealing. Renal ultrasound was unremarkable. His renal function failed to improve with fluids or diuretics. Given the degree of proteinuria, lack of improvement after cessation of vancomycin and absence of other nephrotoxins, he was thought to have post-infectious glomerulonephritis and a kidney biopsy was recommended. Unfortunately he chose to defer. He was discharged on daptomycin and warfarin with serum creatinine of 3.0mg/dL and a plan to continue close outpatient follow-up.

He returned a few weeks later with fatigue and was found to have serum creatinine of 7.2mg/dL. Hemodialysis was initiated due to volume overload and oliguria. It was again recommended to get a renal biopsy as the etiology of kidney injury remained undiagnosed. He agreed. Biopsy revealed an IgA predominant glomerulonephritis, diabetic glomerulosclerosis and tubular necrosis with frequent red blood cell casts consistent with warfarin-induced nephropathy. He was started on steroids and anticoagulation was stopped indefinitely.

Discussion: Anticoagulation-related nephropathy (ARN) is a significant complication of anticoagulation that has been associated with accelerated progression of chronic kidney disease and an increase in all-cause mortality. ARN was originally described in patients who were on warfarin and had distinct findings of glomerular hemorrhage on biopsy. Due to the challenges of biopsy while on therapeutic anticoagulation, this entity may be under-diagnosed. ARN is now defined as acute kidney injury without obvious etiology in the setting of supra-therapeutic INR. Interestingly, our patient did not have a supra-therapeutic INR throughout his course of anticoagulation but biopsy was nonetheless consistent with the diagnosis.

In undifferentiated kidney injury that fails to improve, it is essential to pursue biopsy as it may alter management and prognosis. Unfortunately, the patient was reluctant to undergo biopsy which lead to a delay in diagnosis and subsequent end-stage renal disease. Reflecting on this case, warfarin was started in the setting of an upper extremity superficial venous thrombosis. This also serves as a reminder to weigh the risks and benefits of anticoagulation as side effects are not limited to bleeding.
It’s Never Too Late

Authors: Christina Al Malouf, Martine Al Malouf, Xavier Jimenez

Introduction: We present a case of bacterial meningitis with bacteremia secondary to sphenoidal sinusitis causing contiguous spread to the meninges, due to the presence of a cerebrospinal fluid (CSF) leak caused by retained surgical material from transsphenoidal surgery that was performed over 30 years ago.

Case Presentation: 69 year old male with panhypopituitarism after transsphenoidal resection of a pituitary adenoma followed by radiotherapy performed 35 years ago, on replacement therapy presented to our hospital with a subacute onset of a new headache occurring daily for 1 month and a 48 hour history of rapid altering mental status. On arrival to our hospital, patient was found to be febrile with temperature 101.4 F. On examination, patient was pleasantly confused, without any other abnormalities. CT head was performed in ER which demonstrated an enlarged soft tissue in the sella suspicious for possible recurrent adenoma. Lumbar puncture was performed and fluid analysis was concerning for bacterial meningitis with WBC >3K, neutrophil predominant, very low glucose and elevated protein. Patient received intravenous Vancomycin, Cefepime, Ampicillin, Voriconazole, Acyclovir and Dexamethasone. MRI was unable to be performed due to patient factors. Dedicated CT of the sinuses was subsequently performed revealing a pituitary sellar enhancing 2 cm collection and progressive osseous changes within the sphenoid sinus posterior wall and contiguity with enhancing tissue filling sphenoid sinus. Neurosurgery was consulted and patient was taken to the operating room (OR) for urgent transsphenoidal pituitary drainage and washout with closure of CSF leak. Silastic material presumed to have been from his prior pituitary adenoma resection 35 years ago was retrieved in the OR, which was thought to be the nidus for the CSF leak and presumed cause for contiguous meningitis. OR cultures grew Pseudomonas Aeruginosa sensitive to Cefepime and Methicillin Sensitive Staphylococcus Aureus. Blood cultures drawn on day of presentation also grew Methicillin Sensitive Staphylococcus Aureus, likely haematogenous spread from the sellar abscess. His blood cultures cleared within 24 hours of surgery. Interestingly, his CSF cultures did not grow any organisms. Acid Fast Bacillus cultures were also taken from OR tissue due to subacute presentation which was also negative. Antibiotics, antiviral and antifungals were deescalated accordingly and patient’s neurological status completely resolved. Patient was discharged home without any neurological deficits and completed 6 week course of Cefepime.

Discussion: Persistent CSF leaks are the leading cause of morbidity following transsphenoidal surgery, its incidence remains high at 2.7%. Most of these CSF leaks are observed intra-operatively or within first 1-2 weeks of surgery. To our knowledge, the most delayed leak that has been presented in the literature was within 4 months of surgery. The risk of postoperative CSF leak is significantly increased with prior radiotherapy, due to thinning of the bone at the skull base and predisposing patients to delayed CSF leak.
Is Dialysis really working? A case of pericardial effusion in a patient with End Stage Renal Disease on dialysis.

Authors: Dr. Jessita Albert Messiah Dhas, MD, Dr. Sudheer Penupolu, MD, Dr. Ion Dan Bucaloiu, MD.

Introduction: Uremic pericardial effusion in End stage renal disease (ESRD) is commonly due to inadequate dialytic clearance and non-adherence to dialysis. We present a case of large pericardial effusion in a patient with ESRD who was compliant with scheduled hemodialysis (HD).

Case Presentation: An 81-year-old man, with ESRD due to hypertension on thrice weekly HD for 5 years presented with nausea, vomiting and generalized weakness. He was in septic shock from left lower lobe pneumonia. Computed tomography of the chest revealed a large intermediate density pericardial effusion. Echocardiogram confirmed pericardial effusion without evidence of tamponade. His shock improved with IV antibiotics and he was weaned off pressors. Infection, malignancy, collagen vascular disease, hypothyroidism, myocardial infarction were ruled out as potential causes for his pericardial effusion. Review of records from his outpatient dialysis unit revealed a significant decrease in his dialytic clearance studies. A tentative diagnosis of uremic pericarditis was made. Given the lack of tamponade physiology, a therapeutic pericardiocentesis was not performed. An arteriovenous fistulography revealed a severe stenosis of the cephalic arch vein with thrombus formation in previously placed stents. Balloon angioplasty and thrombectomy were performed with excellent results. After 2 weeks of intensive daily dialysis he had resolution of the effusion.

Discussion: Pericardial effusion is a known complication of uremic syndrome which has become less common since the advent of dialysis. HD circuit recirculation, is a phenomenon in which a mechanical obstruction separates the blood that is dialyzed form the systemic circulation. It can manifest as low dialytic clearance (suboptimal URR or Kt/V), or as mechanical complication of dialysis sessions such as elevation in venous pressures, high negative arterial pressures or prolonged post decannulation bleeding.

With a thorough history, chart review and comprehensive approach involving nephrologists and interventional radiologists, we avoided an unnecessary invasive procedure (pericardiocentesis) that would otherwise have been performed. This case illustrates the need for internists to be diligent in patients who are on HD but have symptoms suggestive of inadequate renal clearance. Absolute values of BUN and creatinine in isolation might not suffice in making this diagnosis and multi-disciplinary approach is in the patient’s best interest.
Life Threatening Complication of Myocardial Infarction: A Case of Myocardial Rupture

Authors: Muhammad Usman Ali, MD, Muhammad Hassaan Sattar, MD, Jonathan Finkel, MD

Introduction: Myocardial rupture is a life threatening complication that usually occurs in the setting of myocardial infarction (MI) or trauma. Myocardial rupture usually involves ventricular free wall, septum or papillary muscle, in decreasing order of frequency with mortality rate of 50% at 5 days. The incidence rates of ventricular septal rupture varies from 0.2%-4.6%. In most cases, clinical presentation occurs within three to five days following acute myocardial infarction. Female sex, age greater than 60 years are risk factors for myocardial rupture while successful percutaneous intervention (PCI) is a protective factor. Currently, incidence and mortality from myocardial rupture is decreasing. We present a case of myocardial rupture after myocardial infarction.

Case Presentation: A 84 year old male with past medical history of three-vessel coronary artery bypass graft surgery, hypertension presented with chest pain. On examination, blood pressure 130/70 mmHG and heart rate 45 beats/min. An EKG showed complete heart block with dynamic ST depressions. A temporary transvenous pacemaker was placed. Troponin trended from <.02 to 5.69 (insert number) and a bedside echocardiography was performed that showed normal left ventricular systolic function with inferior wall hypokinesis. Patient was diagnosed with non-ST elevation MI and heparin drip was started. A cardiac catheterization was performed that revealed a known occluded right coronary artery as well as occluded vein graft to the PDA which was thought to be the culprit for the ACS presentation. After discussion with family, no intervention was done given the high risk of vein graft intervention and the lack of ongoing ischemic symptoms. On day 5, the patient developed acute severe chest pain, and bedside echocardiogram showed pericardial hematoma. A CT scan of the chest was done that revealed defect within the left ventricular free wall, with a communicating rupture and active hemorrhage into the pericardium and a large pericardial hematoma compressing the atria [image]. After lengthy discussion with patient and family about risk versus benefits of surgical intervention, the patient was made comfort care and patient passed away the next day.

Discussion: Our patient had an acute myocardial infarction which eventually lead to myocardial rupture. Our patient was high risk for myocardial rupture due to age and absence of revascularization. Myocardial rupture can present as chest pain, dyspnea, syncope, cardiogenic shock, cardiac tamponade, hemothorax, and sudden death. Bedside echocardiography is the modality of choice for prompt diagnosis and computed tomography/magnetic resonance imaging are reasonable options when in doubt.

Due to the rarity as well as the extremely high mortality rate, it is important to keep a high level of suspicion for myocardial rupture following acute myocardial infarction, especially when prompt revascularization is not performed. Early diagnosis and surgical intervention is crucial to improve survival.

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Docetaxel-induced pneumonitis: An uncommon phenomenon with unusual occurrence

Authors: Zubair S Bashir MD, Syed M Bukhari MD, Fritz Lubin MD

Introduction: Docetaxel is known to cause lung toxicity, although the incidence is very rare. In our case, this difficult diagnosis was challenged by the delayed presentation and drug interaction.

Case Presentation: 85-year-old male with diagnosed history of prostate and urothelial carcinoma presented with new onset dyspnea at rest which worsened on exertion and was associated with night sweats, chills and fever. He was hypoxic, requiring 4 liters through the nasal cannula, which gradually worsened. Enhanced CT chest showed multifocal ground glass opacities with interstitial thickening in septae and fissures, not seen in the CT chest done about two months ago. Blood cultures, sputum analysis and RVP were negative. Blood count, CMP and BNP were unremarkable. He was started on broad-spectrum antibiotics. There was no significant clinical improvement with diuretic therapy. He was also started on steroid therapy. No prior history of radiation exposure to the chest was identified. In retrospect, he had been on weekly therapy with docetaxel for the last 6 months for castration-resistant prostate cancer. In addition, two months ago he was started on amiodarone for his supraventricular tachycardia.

Discussion: The literature review on docetaxel-induced pneumonitis shows the onset of symptoms in approximately 10 to 20 days after docetaxel administration. Our case illustrates the potentially delayed manifestation of docetaxel-induced pneumonitis in the setting of concomitant treatment with amiodarone in a patient who had earlier tolerated many cycles of docetaxel without any side effects. Amiodarone has the potential to increase the levels of docetaxel in serum. Given long half-life of amiodarone, the pneumonitis in our case was secondary to docetaxel toxicity in the setting of drug-to-drug interaction with amiodarone. Docetaxel has the potential to cause lung toxicity. Given the fact that amiodarone has a long half-life and has potentially-fatal drug-drug interaction with Docetaxel, one should apply caution while using the two drugs simultaneously in the clinical setting.

References

What are the ODS? A Case of Osmotic Demyelination Syndrome Associated with Hemodialysis in End-Stage Renal Disease

Authors: Ellen Bradley, PGY-II, Yvette Wang, DO

Introduction: Osmotic demyelination syndrome (ODS) is the irreversible demyelination of the central pons, among other areas of the brain, that is most often associated with rapid correction of hyponatremia. Per contra, reversible ODS is a rare state that can be seen in patients with end-stage renal disease (ESRD) on hemodialysis (HD). This clinical case offers an alternative problem representation for acute altered mental status in a patient with ESRD on HD.

Case Presentation: A 76yo African American female with a past medical history of ESRD on HD, diabetes mellitus, hypertension and hyperlipidemia presented to the ED with new-onset aphasia and confusion. Patient is compliant and completed dialysis that morning. Home medications include glipizide, insulin, amlodipine, metoprolol, and simvastatin. Vitals at presentation were within normal limits except a blood pressure of 195/86. At admission, the patient exhibited global aphasia and apraxia, but moved all four extremities spontaneously. GCS was 9 and NIHSS was 7, later increasing to 14. Hemoglobin was 12.7, and sodium was 138. Recent outpatient labs were normal. CT head, CT angiogram, and CT perfusion did not show acute intracranial ischemia or hemorrhage. However, MRI brain showed diffuse increased T2 signal intensity within the pons, suggestive of osmotic demyelination. Aspirin 325mg was administered, and aspirin and high-intensity statin were continued for concern of cerebral vascular accident (CVA). EEG was negative for seizure. There were no active signs of infectious or autoimmune etiology. Urine drug screen was not performed due to anuria. Repeat MRI and MRA brain/head/neck was negative for CVA or reversible posterior leukoencephalopathy syndrome, but continued to show osmotic demyelination in the pons. Sodium remained between 135 and 141. Supportive treatment was initiated without antibiotics or steroids: blood pressure was kept normotensive, electrolytes were kept within normal limits, and fluid shifts were minimized during HD. The patient spontaneously recovered, and at discharge six days later, she was at her neurologic and cognitive baseline. Aspirin was discontinued, and home statin was continued.

Discussion: Rapid correction of hyponatremia >10-12mEq in 24 hours is the most common cause of ODS, but case reports have also shown ODS in patients undergoing dialysis; these case reports correlate with our patient with global aphasia and apraxia. Treatment of this type of ODS consists of minimizing fluid shifts during HD and maintaining normotension. The patient had three HD sessions in her hospital stay, two of which were filtration without fluid exchange. Extensive workup was negative for other etiologies, and symptoms resolved without other intervention. Although rare, HD-associated ODS should be considered in the problem representation of altered mental status in patients with ESRD in the right clinical context. It is imperative that more research is conducted on its cause and prevention in the future.
Metastatic Melanoma to the Stomach: A Rare Cause of Upper Gastrointestinal Bleeding

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Introduction: Upper gastrointestinal (GI) bleeding is a common diagnosis with annual incidence of hospitalization reported as 100 per 100,000 individuals and etiologies are many. The most common causes are generally recognized to be esophageal varices or peptic ulcer disease. Neoplasm in the stomach is a less common cause, and is usually due to primary gastric adenocarcinoma. The gastrointestinal tract can also be a site of metastases, however malignant melanoma is a relatively rare cause of upper gastrointestinal bleeding, accounting for less than 3% of all severe cases of bleeding.

Case Presentation: The patient is a 76-year-old male with a history of coronary artery disease status post drug-eluting stent, ischemic cardiomyopathy, hyperlipidemia and melanoma of the face treated thirty years prior, who presented to the emergency department (ED) with hematemesis and melena for 2-3 days prior to admission along with progressively worsening dyspnea on exertion and fatigue. The patient had recently started aspirin and clopidogrel following drug eluting stent placement. Upon evaluation in the ED, the patient was found to have blood pressure 119/78, heart rate 98, respiration rate 18, oxygen saturation 97% on 2 liters via nasal cannula. A physical exam revealed pallor with poor capillary refill. There was no abdominal tenderness. Lab work revealed hemoglobin 5.6, white blood cells 13.3, serum creatinine 1.35, troponin 0.26 à 0.37. In the ED, the patient was given two units of packed red blood cells (pRBC) and intravenous fluids for volume resuscitation and repeat blood work showed a hemoglobin of 7.6. The patient was evaluated by gastroenterology who recommended esophagogastroduodenoscopy (EGD). The patient was admitted to low level monitoring for continued blood transfusion requirement.

The patient was initially managed with volume resuscitation using crystalloids and blood products. EGD showed large gastric ulcer with heaped margins along the greater curvature of the stomach with a red the center. An endo clip was placed for hemostasis. Biopsies were obtained which later revealed to be metastatic malignant melanoma with S100 and MART1 positivity. Next-generation sequencing was performed demonstrating negativity for BRAF, NRAS, and KIT mutations. Computed tomography of the head showed an enhancing spherical lesion in the right basal ganglia concerning for metastatic disease.

Discussion: Patients with metastatic melanoma to the GI tract have an overall poor prognosis with life expectancy as short as four months. With the rising incidence of melanoma and its subsequent GI metastasis, it is important to recognize metastatic disease early and potentially decrease the chance of life-threatening complications, which may lead to an early death. Only approximately 4% of patients with gastrointestinal melanoma are diagnosed prior to death. Prophylactic evaluation with endoscopy may be a powerful detection technique in patients diagnosed with melanoma and could detect early disease and provide treatment options to patients.

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Psoriasis as the single risk factor for “Locked-in syndrome” after ischemic stroke in a young patient

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Introduction: Locked-in syndrome (LIS), also known as cerebromedullospinal disconnection or pseudocoma, is a rare neurological disorder. LIS is most commonly caused by ischemic or hemorrhagic pontine stroke. Other causes such as trauma, infection, tumors, demyelinating disease and amyotrophic lateral sclerosis may also cause LIS. Autoimmune diseases have been shown to increase risk of cardiovascular disease and cerebrovascular disease. Psoriasis as an isolated risk factor for LIS has not been described in literature

Case Presentation: A 50-year-old male patient with psoriasis (for which he was on no treatment) presented to the ER after being found in his home with loss of consciousness. He had no other medical problems. Admission blood tests and drug screen were negative. Initial CT angiogram of head and neck showed acute complete occlusion with thrombus of the middle and distal basilar artery truck. He was intubated and admitted to the intensive care unit. Patient had mechanical thrombectomy of basilar artery truck and right superior cerebellar artery with radiological restoration of normal brain perfusion from TICI (Thrombolysis in Cerebral Infarction) Score 0 to TICI 3. However he did not clinically improve and continues to have respiratory failure, dysarthria, dysphagia, and quadriplegia. Patient is able to communicate only via a rudimentary system by blinking his eyes for yes or no answers. He had tracheostomy and was transferred to a long-term acute care hospital.

Discussion: Based on our case and published reports, psoriasis patients are known to have higher risk for stroke over general population. Several meta-analyses have quantified this risk; for patients with mild psoriasis, the risk ratio (RR) for stroke was 1.12 (95% CI, 1.08-1.16) and for patients with severe psoriasis, the RR for stroke was 1.56 (95% CI, 1.32-1.84). The mechanism for association between psoriasis and stroke is unclear. Systemic inflammation is the most likely mechanism by accelerating atherosclerosis via inflammation-induced endothelial dysfunction. Supporting this hypothesis is the fact that psoriasis is associated with increase in morbidity and mortality from all forms of cardiovascular diseases. Studies have pointed out that psoriasis should be considered as a modifiable risk factor for cardiovascular and cerebrovascular diseases.
Diffuse Large B Cell Lymphoma mimicking ST elevation myocardial infarction

Authors: Amir Eslami, DO; Osama Mahmoud, MD; Jeffrey Rickert, MD; Carl Shultz, DO; Amro Alsaid, MD.

Introduction: Primary cardiac lymphoma is rare and difficult to diagnose due to nonspecific symptoms. Secondary infiltration is more commonly seen in 25% of patients with disseminated disease, most notably in the immunocompromised population. Typically, these patients present with dyspnea on exertion, angina, atrioventricular (AV) block, and constitutional symptoms. We present a case of a 59-year-old immunocompetent man who presented with rapidly progressive cough found to have ischemic changes on his electrocardiogram (ECG).

Case Presentation: A 59-year-old Caucasian man with a past medical history of hypertension, insulin-dependent diabetes, tobacco use, and microcytic anemia presented to the emergency department with the chief complaint of progressively worsening non-productive cough for two months. He endorsed dyspnea on exertion, fatigability, and a 10-pound weight loss. Physical exam was remarkable for hemodynamic stability, diminished bibasilar lung sounds, and central obesity. Laboratory results revealed a hemoglobin of 13.0 g/dL, MCV of 77.4 fL, platelets of 626 K/μL, BUN of 24 mg/dL, Creatinine of 1.5 mg/dL, calcium of 14.0 mg/dL, and pro-BNP of 4577 pg/mL. Cardiac troponin T by high sensitivity assay was increased at 124 ng/L with a repeat of 119 ng/L. An ECG revealed ST segment elevations in leads II, III, aVF with Q-waves and reciprocal ST depressions in leads I and aVL. He was given aspirin and started on intravenous unfractionated heparin. A non-contrast chest CT scan was remarkable for cardiomegaly, large pericardial effusion, diffuse soft tissue nodules and thickening in the pericardium, and a confluent mediastinal, right hilar, perihilar and left hilar lymphadenopathy. The transthoracic echocardiogram (TTE) showed right ventricular and bi-atrial enlargement with thickening in the AV groove between the right atrium and ventricle. Cardiac magnetic resonance imaging (MRI) showed multiple mass-like lesions within the pericardium, the largest measuring 63 mm x 40 mm encasing the right coronary artery and evidence of myocardial infiltration with increased T2 values. Patient underwent a cardiac biopsy, which revealed mediastinal diffuse large B cell lymphoma (DLBCL). PET scan showed markedly hypermetabolic lymphoma involvement in the mediastinum and pericardium. The patient was started on chemotherapy with dose-adjusted R-EPOCH with plans for six cycles of therapy. After three cycles of chemotherapy, a repeat PET scan showed near complete resolution of pericardial and myocardial abnormal metabolic activity.

Discussion: This case demonstrates an atypical presentation of a rare disease. Despite multiple coronary artery disease risk factors, physicians must be aware of alternative diagnoses in patients with atypical symptoms and ischemic ECG changes. It illustrates the importance of multimodal cardiac imaging in early detection, rapid diagnosis, and safe treatment of myocardial infiltrates secondary to DLBCL. Furthermore, it confirms that standard therapy for DLBCL can be effective with primary cardiac involvement and minimal adverse events.

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Hemihyperplasia and Deep Vein Thrombosis: Clues to a Rare Underlying Syndrome

Authors: Tamaryn Fox, MD, Zurab Azmaiparashvili, MD, Peter Moussa, MD, Jaber Monla-Hassan, MD, FCCP

Introduction: Klippel-Trenaunay syndrome is a rare congenital condition characterized by a clinical diagnosis fulfilling the triad of capillary malformations (manifesting as port-wine stains), venous malformations and unilateral overgrowth of bone and/or soft tissue of the extremities. Genetically, it usually occurs as sporadic, somatic mutations; however, rarely germline mutations may lead to a para-dominant familial pattern.

Case Presentation: A 68 year old female patient presented with a history of worsening swelling of her right lower extremity for a month duration, associated with pain and paresthesias. On past medical history she noted having had multiple deep vein thromboses first occurring at age 19. She was subsequently put on warfarin. An inferior vena cava (IVC) filter was placed when the clots continued to propagate despite anticoagulation therapy. She did not remember ever being told the reason for her recurrent clots. She did state that since birth, her right side has been bigger than the left. She also noted that her father had the same condition; however his left side was bigger than the right.

On examination the patient was found to have a port-wine stain of her right upper arm extending into her axilla and hemihyperplasia of the right upper and lower extremity. Her right lower limb showed early signs of phlegmasia cerulea dolens with some blueish discoloration and marked swelling.

Ultrasound doppler imaging of bilateral lower extremities (2013) noted that there was a deep vein abnormality which could be congenital in nature.

CT angiogram of the chest with venous runoff performed and no pulmonary emboli were identified, however did show on the venous runoff phase, extensive deep vein thrombosis extending down bilaterally from the level of the inferior vena cava, and also demonstrated right lower extremity varicose veins.

She underwent three extensive interventional radiology procedures: catheter directed tissue plasminogen activator (TPA), suction thrombectomy and recanalization of the IVC with stent placement. She was put on a heparin infusion peripherally and will be bridged to oral anticoagulation therapy.

Discussion: Complications related to Klippel-Trenaunay syndrome include a predilection for the development of deep vein thromboses in view of the underlying venous malformations, bleeding, recurrent cellulitis and lymphedema.

Treatment is mainly supportive. Treatment modalities do exist for the vascular malformations. Sclerotherapy, laser, embolization and sirolimus for extensive disease have all been described. From a medical standpoint, treatment includes managing complications such as anticoagulation for recurrent DVTs, treatment of recurrent cellulitis and management of pain.

Patients with Klippel-Trenaunay syndrome tend to present with severe complications. By early recognition of this syndrome, one can involve a multidisciplinary team and establish continued care and monitoring to try and prevent severe complications from occurring such as extensive, severe, limb-threatening deep vein thrombosis, pulmonary embolism, recurrent cellulitis and chronic debilitating lymphedema.

References
TAFRO Syndrome: A Rare Manifestation of Multicentric Castleman’s Disease

Authors: Elizabeth Gold, DO; Ross Biggs, DO

Introduction: Thrombocytopenia, anasarca, fever, renal dysfunction, and organomegaly (TAFRO) Syndrome is a rare variant of Multicentric Castleman’s Disease (MCD). It was first described in 2008 with approximately 50 cases worldwide. The diagnostic criteria are histopathology compatible with MCD without HHV-8 infection. TAFRO Syndrome is further delineated from MCD by hyperplastic megakaryocytes, elevated alkaline phosphatase, and absence of hypergammaglobulinemia.

Case Presentation: A 47-year-old male presented with 6 days of progressive cough, shortness of breath, fever, and myalgias. He was admitted for bilateral lower lobe pneumonia and placed on antibiotics. Despite antibiotic therapy, his respiratory status worsened requiring intubation. Work up included a CBC, CMP, and CT scan of the chest, abdomen, and pelvis. CT scan was notable for ground glass opacification within both lung bases, small bilateral effusions, cardiomegaly, multiple areas of lymphadenopathy, and ascites. Laboratory evaluation revealed thrombocytopenia, anemia, leukocytosis, elevated alkaline phosphatase, and acute renal failure. A lymph node core needle biopsy came back negative for lymphoproliferative disorders. A bone marrow biopsy was performed which showed megakaryocytic hyperplasia. Further evaluation for infectious etiologies with a respiratory viral panel, lumbar puncture, bronchial-alveolar lavage, blood cultures, tick-borne illness serologies, HIV, and EBV/CMV serologies came back negative. Immunologic work up was notable for a mild increase in IL-6 level at 6, but negative for IgG abnormalities or hypergammaglobulinemia. Definitive diagnosis was established with excisional lymph node biopsy, which revealed features most consistent with MCD in the setting of TAFRO syndrome. The patient was started on Siltuximab with improvement in his clinical status.

Discussion: Since TAFRO syndrome is a rare manifestation of MCD, there is still more research that needs to be done. In 2015, diagnostic criteria were set forth to facilitate the diagnosis of TAFRO syndrome. They were separated into major and minor criteria. The three major criteria are thrombocytopenia, anasarca, and systemic inflammation. The four minor criteria are lymph node histopathological features of MCD, bone marrow myelofibrosis and/or hyperplasia of megakaryocytes, organomegaly, and renal insufficiency. TAFRO syndrome should be considered if three major criteria and two minor criteria are present. The patient in our case had all major and minor criteria present.
A Fatal Case of Severe Babesiosis Presenting as Fulminant Liver Failure

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Introduction: Babesiosis is a tick-borne zoonotic infection caused by the protozoan Babesia. Transmitted by the Ixodes tick, B. microti is endemic to the Northeastern and Midwest United States. The initial presentation typically includes flu-like symptoms, fever, chills, myalgias, and headaches. Immunocompromised and older individuals are at risk of developing life-threatening complications, including multi-organ failure and disseminated intravascular coagulation. Rapid diagnosis is imperative to identify patients at risk of severe complications and initiate effective antimicrobial therapy.

Case Presentation: A 79-year-old male from New Jersey with a past medical history significant for end-stage renal disease, coronary artery disease with coronary artery bypass grafting and hypertension, was transferred to our institution for evaluation of significantly abnormal liver function tests concerning for drug-induced liver injury. Two weeks prior to his presentation, the patient had flu-like symptoms and began a 5-day course of amoxicillin-clavulanate 500mg twice daily. His symptoms persisted and was taken to his local hospital. Laboratory tests showed alanine aminotransferase (ALT) and aspartate aminotransferase (AST) levels of 2,335IU/L and 3,710IU/L, respectively; and total bilirubin of 3.3 mg/dL. Computed tomographic images of the abdomen revealed splenomegaly and free fluid in the upper abdomen and pelvis. The patient was transferred to our institution’s hepatology service for specialized care and assessment. Additional history revealed no recent travel, history of alcohol use, intravenous drug abuse, or any family history of liver pathologies. He lives at home with three dogs and had no other recent animal contact. Repeat laboratory tests showed rising levels of ALT (2,718IU/L), AST (3,497IU/L) and total bilirubin (5.2 mg/dL; direct bilirubin 3.6 mg/dL). Hemoglobin was 7.8 g/dl, white blood cell count of 17.3 g/dl, INR 4.1, and a hepatitis viral panel was negative. A peripheral blood smear (PBS) revealed multiple intraerythrocytic trophozoites with Maltese-cross appearing merozoites, pathognomonic of babesiosis. With diagnostic confirmation, infectious disease was consulted, and the patient was transferred to the intensive care unit. Atovaquone 750 mg twice daily and azithromycin 500 mg once daily were started along with emergent exchange transfusion. Unfortunately, the patient continued to decompensate requiring intubation and vasopressor support for hypotension. On the third hospitalization day, an electrocardiogram demonstrated ST-segment elevations in V1-V2 and T-wave inversions in lead V6, with associated troponin level to 38 ng/mL, indicative of an acute myocardial infarction. The patient died shortly thereafter.

Discussion: We report the case of an elderly male with multi-organ failure secondary to severe babesiosis. The initial impression was drug-induced liver injury due to recent antibiotic use. However, the associated hematological abnormalities led us to consider other diagnostic possibilities. A PBS should be considered in all patients presenting with acute liver failure and a strong epidemiologic history for babesiosis. Once suspected, prompt and appropriate treatment must be initiated to reduce the morbidity and mortality from this infection.
An Uncommon Cerebrovascular Pattern of Stroke in an Asian patient

Authors: Ronan Hsieh, MD, Jessica Stempel Velasco, MD, Saman Zafar, MD, Akanksha Agrawal, MD, Pradhum Ram, MD, Jorge Penalver, MD, Andres Mora, MD, Mario Naranjo, MD

Introduction: Moyamoya disease is a chronic progressive cerebrovascular disease more commonly seen in Asian population. We present a case of Moyamoya disease with pathognomonic neuroimaging findings.

Case Presentation: A 34-year-old Asian woman with history of anxiety, hyperthyroidism and hypertension, presented with confusion, progressive right upper and lower extremity weakness and paresthesia for four days. She complained of worsening short-term memory loss and ambulatory dysfunction. She denied fever, chills, neck rigidity, recent trauma or vision changes. Her vital signs were stable without fever. On physical exam, she had right nasolabial fold flattening, and right upper and lower extremity muscle strength of 4/5. Her high cortical functions, vision field, sensory exam, reflexes and left-sided muscle strength were intact. Her findings were consistent with a National Institutes of Health Stroke Scale 3. No significant cardiac murmurs or carotid bruits were heard. Laboratory data were unremarkable except for a positive urine drug test for opiates and cocaine. An electrocardiogram showed regular sinus rhythm. Computed tomographic imaging of the head showed a subacute infarct in the left parietal lobe and chronic infarct in the left frontal lobe. Cranial angiogram showed high-grade stenosis with abrupt cut-off at bilateral anterior, middle and posterior cerebral arteries near the skull base, with prominent collateral arteries along the course of lenticulostriate. The angiogram finding was pathognomonic for Moyamoya disease. After stabilization, she was discharged with the plan for internal carotid artery-external carotid artery bypass.

Discussion: Moyamoya disease is characterized by bilateral arterial stenosis around the Circle of Willis [1]. It has some familiar patterns and involves mutation of RNF213 gene on chromosome 17q25.3 [2], although other genetic abnormalities are also possible. Moyamoya disease can affect patients of all ages but is predominantly diagnosed in teenagers and patients aged 40-49 years [3]. Clinical presentations include transient ischemic attack, ischemic or hemorrhagic stroke, headache or seizures [4, 5]. Neuroimaging is usually pathognomonic with bilateral arterial stenosis [6]. Based on the cranial angiogram, Suzuki et al. described the six stages of progress from isolated involvement of carotid fork to the complete obliteration of the internal carotid arteries [1]. Because Moyamoya disease is chronic and progressive, collateral arteries are often prominent. Treatment options for Moyamoya disease include short-term and long-term treatments. The immediate treatment is similar to the management of an acute stroke [7], which can include symptomatic management, hematoma evacuation for hemorrhagic stroke, seizure prevention, and intracranial pressure reduction. Aspirin is recommended for all patients unless contraindicated [8]. Patients with an ischemic cerebrovascular presentation, cognitive decline or progressive symptoms should be evaluated for revascularization surgery [9]. Anticoagulants are currently not recommended in patients with Moyamoya disease because the risk of hemorrhagic stroke is high and there is insufficient evidence demonstrating a direct benefit of anticoagulant use [8].

References


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Nausea and Vomiting in Breast Cancer- Not Your Usual Suspects- An Extremely Rare Case Presentation

Authors: Shumona Ima, MD, Mariette Austin, PhD MD, Mahesh Krishnamurthy, MD

Introduction: Nausea and vomiting are very common symptoms in cancer patients- sometimes due to the cancer itself and at other times as a consequence of the cancer treatments. Gastric outlet obstruction presents with nausea & vomiting – with most common etiologies as ulcer disease or local malignancies. Retroperitoneal fibrosis caused by metastatic invasive lobular breast carcinoma, presenting with nausea and vomiting due to both gastro-duodenal obstruction and hydronephrosis is extremely rare.

Case Presentation: A 76 year old Caucasian female with long-standing history of metastatic breast cancer on chemotherapy presented with progressive upper abdominal discomfort with oral intake, nausea and vomiting. CT scan of her abdomen showed amorphous poorly definable right retroperitoneal soft tissue thickening involving the duodenum, hepatic flexure of the colon and extending to involve the proximal to mid right ureter resulting in severe right hydronephrosis. In addition, the CT showed duodenal narrowing with gastric distention and intrahepatic biliary and pancreatic ductal dilatation. Her PET scan showed insignificant SUV in the soft tissue, felt to be unlikely due to her known and very PET-avid breast cancer. Subsequently, she underwent EGD and EUS with FNA twice prior to being diagnosed with metastatic breast cancer, by means of immunohistochemistry analyses. She also underwent duodenal and ureteral stent and PTC placements for symptomatic relief.

Discussion: This case is unique because both retroperitoneal fibrosis and gastric outlet obstruction are exceedingly uncommon presentations for metastatic breast cancer. Moreover, PET scan and initial fine needle aspiration biopsy did not show enough evidence to support retroperitoneal metastasis of breast cancer. Extensive literature review revealed that metastatic breast cancer is only rarely diagnosed as a result of an initial presentation with gastric outlet obstruction or retroperitoneal fibrosis with hydronephrosis. However, malignancy should always be definitively ruled out before drawing any diagnostic conclusions. Immunohistochemical analyses of biopsy specimens represent the most informative mechanism for diagnosing metastatic carcinoma.

Conclusion: Identifying retroperitoneal fibrosis as a cause of nausea and vomiting is helpful, as medical treatment cannot help the gastric outlet obstruction in these cases- for obvious reasons. Needless to say, the prognosis for these patients is very poor. It is important to search for other potential serious causes of nausea and vomiting in cancer patients.

References

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Introduction: It is estimated that acute myocardial infarctions complicate 1 in 16,000 pregnancies in the United States. Up to 25% of these are due to Pregnancy-associated Spontaneous Coronary Artery Dissections (p-SCAD), which have high mortality rates. Majority of these cases have been reported in the postpartum period, typically one week after delivery. Very few dissections have been reported in the antepartum period, almost none involving the entire left anterior descending artery. Conservative management with beta blockers is the mainstay of treatment in stable p-SCAD. We present a case of unstable p-SCAD with cardiogenic shock treated with percutaneous coronary intervention (PCI).

Case Presentation: A 36-year-old G3/P1102 with twin gestation was scheduled for an elective cesarean section for breach and transverse lie. Past medical history was significant for migraine and previous cesarean sections. She was a non-smoker and did not carry any personal or family history of risk factors for coronary artery disease. At 37 weeks’ gestation, she presented with acute substernal non-radiating chest pressure, shortness of breath, nausea, and diaphoresis. Her vitals and physical exam on presentation were essentially unremarkable. Electrocardiogram showed acute anterolateral ST-elevation myocardial infarction (STEMI). Labs showed hemoglobin (Hb) of 7 gm/dl (baseline 12.3 gm/dL). Emergent cardiac catheterization revealed spontaneous dissection of the left anterior descending artery extending from proximal to distal vessel, severe anteroapical and anterolateral hypokinesis with an estimated ejection fraction (EF) between 20-30%. Due to the extent of dissection, coronary artery bypass grafting was not deemed feasible by the cardiothoracic surgery team. Seven overlapping drug-eluting stents were placed following which her EF improved to 35-40%. Her catheterization was followed by a successful emergent cesarean section and delivery. Her post-operative ICU course was complicated with hemodynamic instability requiring pressor support. Hb was 6 gm/dl, and she was transfused a total of 6 units of packed red blood cells. Repeat ECHO showed decreased EF of 25-30% with severe apical and entire anterior wall hypokinesis. She was placed on an intra-aortic balloon pump (IABP). She continued to have heavy lochia in the post-operative period for which coil embolization of uterine arteries was done. The balloon pump was removed after two days. She was managed with Ramipril, Carvedilol, and Spironolactone once she was stabilized. EF on hospital day six improved to 45%. She was discharged home with cardiac rehab and close follow-up.

Discussion: Our case adds to the present knowledge of complications associated with p-SCAD. Possible associations with hemodynamic stressors, oxytocin release in breastfeeding mothers, older/multiparous mothers, relationship to eclampsia/pre-eclampsia, peripartum cardiomyopathy, fibromuscular dysplasia, and other extra-coronary vascular abnormalities have been implicated but need further research.

References

Cryptococcal Meningitis in A Patient with Chronic Lymphocytic Leukemia on Ibrutinib; An Emerging Risk Factor for Invasive Fungal Infections.

Authors: Noman Ahmed Jang Khan, MD, Sheetal Higbee, MD, Hassan Mehmood, MD, Rai Shahjehan Dilawar, MD

Introduction: Cryptococcus is a unique environmental fungus that often affects immunocompromised individuals with abnormal cell mediated immunity like Human Immunodeficiency Virus (HIV) [1]. Other known risk factors include solid organ or bone marrow transplant patients, patients receiving immunosuppressive agents, patients with very advanced malignancies. Very few cases of cryptococcal disease have been reported in patients with Chronic Lymphocytic Leukemia taking Ibrutinib, a newer biological agent primarily affecting humoral immunity by inhibiting the Bruton’s Kinase Pathway and has shown improvement in the Progression Free Survival in recent studies [2]. We here report a case of cryptococcal meningitis in a patient with CLL on Ibrutinib therapy.

Case Presentation: 79-year-old female came to the Emergency department with complains of headache, fever and malaise. Her past medical history is significant for Diabetes Mellitus type 2, Chronic Lymphocytic Leukemia (CLL) and hypertension. Medications included metformin, amlodipine and Ibrutinib started recently. Emergent Lumbar Puncture was planned, and patient was started empirically on Vancomycin, Ceftriaxone and Ampicillin. Cerebrospinal fluid analysis (CSF) came back remarkable for WBC count of 296 with 31% lymphocytes, glucose low at 35, protein elevated at 120, negative HSV PCR and positive Cryptococcal antigen with a titer of > 1:40. She was started on Liposomal Amphotericin B with IV Flucytosine. CSF culture eventually grew Cryptococcus neoformans susceptible to Amphotericin B [Figure 1]. Patient showed significant improvement in her symptoms and repeat CSF analysis in 2 weeks was unremarkable with negative Cryptococcus titers and culture. Patient is currently on maintenance dose of Fluconazole.

Discussion: Cryptococcal meningitis usually presents indolently over a period of a week or two with fever, malaise, headache and occasionally cranial neuropathies [3]. The presence of cough, skin rash and dyspnea suggest disseminated disease. The definitive diagnosis of cryptococcal meningoencephalitis is made by culture of the organism from the cerebrospinal fluid. The presence of cryptococcal polysaccharide antigen titers are strongly suggestive of infection very well before the cultures become positive particularly in high risk patients [4]. Induction therapy with Liposomal Amphotericin B and Flucytosine followed by maintenance therapy with fluconazole is the standard of care.

Cryptococcal meningoencephalitis should be part of the initial differential diagnosis in patients with CLL taking Ibrutinib with suggestive clinical features. Prompt diagnosis and early treatment could prevent fatal consequences.

References

Complete heart block due to intracardiac metastases of squamous cell carcinoma

Authors: Sugandha Singh Landy, MD

Introduction: Secondary cardiac involvement in metastatic lung cancer often goes undetected due to the absence of early symptoms. When the cardiac conduction system has been infiltrated, the presentation may include arrhythmias, including complete atrioventricular block.

Case Presentation: A 52 year-old male with metastatic primary bronchogenic squamous cell carcinoma, status post 2 cycles of gemcitabine/carboplatin, presented with typical chest pain. Initial electrocardiogram showed sinus bradycardia with 2nd degree AV block, ST elevations in the lateral leads, and ST depressions in the anterolateral leads. Initial troponin was negative. He underwent emergent left heart catheterization, which showed no significant coronary artery disease or wall motion abnormality. He was admitted for further work up and management of his chest pain.

He underwent transthoracic echocardiogram, which revealed normal four-chamber size, wall motion, ejection fraction (60-65%), and diastolic filling pattern. However, a large 3 by 4 cm solid tissue mass was seen at the level of the tricuspid annulus, appearing to emanate from the coronary sinus. The remainder of the echocardiogram findings were without significant findings, aside from trace mitral, tricuspid, and pulmonic valve regurgitation. CT chest angiography was performed, which showed hypoattenuating masses inseparable from the lateral left ventricular wall in the inferior right atrioventricular groove, and appeared to have enlarged from previous imaging. These were felt to represent cardiac metastases with intra-cavitary cardiac extension of both left and right heart. PET scan then revealed these masses were FDG-avid, consistent with cardiac metastases.

He was initiated on Paclitaxel. He had a witnessed sudden-onset loss of consciousness, followed by loss of palpable pulse. Chest compressions were performed and two defibrillator shocks were administered with return of pulse. An electrocardiogram revealed complete heart block. Dopamine infusion was initiated in an effort to increase the heart rate, and thus prevent bradycardia-induced ventricular tachycardia. The following morning, however, he had an episode of sustained ventricular tachycardia at >230 bpm requiring CPR and one external defibrillation with ROSC. Electrocardiogram following this event again showed complete heart block, with PVCs and a prolonged QTc.

Following discussions with palliative care team, the patient declined placement of implantable cardiac defibrillator given high risk associated with the procedure due to location of intracardiac metastases. He instead chose to be discharged home with an external wearable cardiac defibrillator (LifeVest). He passed following a ventricular tachycardia cardiac arrest at home.

Discussion: The combination of atrioventricular dissociation on electrocardiogram and FDG-avid intra-cardiac masses near structures crucial for the cardiac conduction system on PET/CT were consistent with cardiac metastases of primary bronchogenic squamous cell carcinoma in this case. The clinical manifestation of secondary tumors located in the myocardium is likely proportional to the degree of myocardial infiltration and invasion into the conduction system.
Interstitial Pneumonia with Autoimmune Features and Catastrophic Antiphospholipid Syndrome Requiring Prolonged ECMO

Authors: Christopher Lenivy DO, Nathan Brewster DO, Daniel Schwed-Lustgarten MD

Introduction: Interstitial pneumonia with autoimmune features (IPAF) is a classification of lung disease involving an interstitial pneumonia coupled with serologic and morphologic findings suggestive of but not specific to any connective tissue disease (CTD). It is unclear how much these patients benefit from immunosuppression beyond corticosteroids; and there is a paucity of cases addressing the outcomes of patients requiring extracorporeal membrane oxygenation (ECMO). Here we present a case of acute hypoxic respiratory failure secondary to IPAF complicated by catastrophic antiphospholipid syndrome (CAPS).

Case Presentation: The patient is a 62 year-old male with history of hypertension and Raynaud phenomenon diagnosed 2 weeks prior to admission. He presents with one week of dyspnea and hypoxia with diffuse ground glass and interstitial infiltrates and consolidation in the lower lobes. Over the following two days he decompensated with worsening hypoxia requiring mechanical ventilation but eventually required salvage VV-ECMO. His course was complicated by thrombosis in multiple extremities, digital ischemia and subsegmental PEs. The autoimmune panel showed high-titer anti-β2-GP1 IgM, anti-cardiolipin, and lupus anticoagulant. Treatment included a pulse of corticosteroids, cyclophosphamide, and rituximab as well as plasma exchange. After 35 days of ECMO he was successfully decannulated. He received a tracheostomy and PEG tube and was successfully transitioned to collar on day 61 and discharged to LTAC on day 79. The current plan is continuation of hydroxychloroquine, prolonged steroid taper and transition to azathioprine, and warfarin.

Discussion: A corticosteroid pulse with prolonged taper transitioned to a steroid-sparing agent has historically been used for patients categorized with IPAF. This patient with IPAF complicated by CAPS responded to a combination of corticosteroids, hydroxychloroquine, cyclophosphamide, rituximab and plasma exchange while being supported on VV-ECMO.

IPAF is a complex and poorly understood entity. This case of a man with IPAF and multiple life-threatening complications emphasizes the need to utilize a multidisciplinary team of specialists to tackle such a case. Future studies looking at progression to CTD, long-term survival rates, as well as tailored treatment courses will be an important next step.

References

Melanomas Masquerading as Lipomas, and the Morbid Consequences

Authors: Matthew Lipinski, DO; Diane Hershock, MD, PhD

Introduction: Skin lesions are encountered very frequently by internists, and are a common reason for patients to make a doctor’s appointment. Melanomas can have multiple different colors, or be non-pigmented and resemble subcutaneous nodules. The "ABCDEs" of melanoma (asymmetry, border irregularity, color abnormalities, large diameter, enlarging rapidly) can be applied to any concerning skin lesion. Any rapidly growing skin lesion warrants a discussion with the patient about potential biopsy.

Case Presentation: A 22 year-old woman presented with several months of multiple progressively enlarging subcutaneous nodules. Over the past week she had also developed diplopia and nausea. These nodules had quadrupled in size over the past several months. She does not go outside often, and uses sunscreen when she does. Two siblings have had biopsy-proven lipomas in the past. There is no history of skin cancer in her family. Small flesh-colored, freely mobile nodules with irregular borders were present on her right axilla, left arm, back, and chest wall. There was a large 5 cm flesh-colored nodule on her right scalp. There were two 1 cm blue-pigmented nodules in her left axilla. Skin biopsy of a left arm nodule revealed metastatic melanoma. MRI of her brain revealed a 4 cm lesion in the right occipital lobe with radiographic signs of metastatic melanoma.

Discussion: The most common type of melanoma is superficial spreading, which presents as intensely dark, macular structures with irregular borders on the skin surface. However, melanomas can have multiple different colors, or be non-pigmented and resemble subcutaneous nodules. Nodular melanoma, especially when skin-colored, usually has the worst prognosis, partly due to delay in recognition. Any skin lesion with concerning features ("ABCDEs") warrants discussion about possible biopsy. Only about 10% of melanomas are familial-related, so lack of familial skin cancer is not enough to quell suspicions. Metastatic melanoma portends an incredibly poor prognosis, and can present in a wide array of patterns. Multiple palpated or radiographically-identified subcutaneous nodules should raise significant concern. Although there are syndromes that cause multiple lipomas, such as Dercum disease and familial multiple lipomatosis, clinicians should be wary of any skin lesion that grows rapidly. Treatments have evolved from the prior poorly efficacious chemotherapy, and now center around immunotherapy with CTLA-4 inhibitors and PD-1 receptor inhibitors. BRAF mutations allow for treatment targeted toward the B-Raf kinase. Despite these advances, the overall average prognosis is still approximately two years. Due to its many possible clinical presentations and high morbidity and mortality, internists and hospitalists need to have a high clinical suspicion for melanoma when presented with abnormal skin findings. Treatment options are improved but overall survival is still very poor. Thus, active vigilance about any concerning skin lesions, no matter the lesion’s shape or color, is the best thing we can do to help our patients.

References


Strongyloides Hyperinfection Syndrome

Authors: Izza Mir DO, Rashmi Dhital MD, Brian Chwieko MD, Frank Liu MD, David Young MD

Introduction: Strongyloidiasis is a parasitic infection endemic to tropical areas, manifesting commonly as asymptomatic eosinophilia or mild chronic gastrointestinal, cutaneous, or pulmonary symptoms. However, in immunocompromised hosts, the disease can disseminate resulting in high mortality, making it important for clinicians to recognize this clinical entity.

Case Presentation: A 70-year-old man with history of heart failure, Crohn’s Disease on chronic steroids, adrenal insufficiency secondary to steroid use presented with confusion and diarrhea. On exam, he was noted to have coarse breath sounds and bilateral lower extremity edema. Chest x-ray was unremarkable. Labs were remarkable for hyponatremia (sodium 122 Meq/L) and anemia (hemoglobin 8.5 g/dL) but no leukocytosis or eosinophilia (WBC 7.2 10E3/Ul, eosinophil 0.020 10E3/uL or 1.6%). Patient’s home diuretics were held, and he was treated with hypertonic saline for symptomatic hyponatremia. The next day, the patient developed abdominal distension with abdominal x-ray showing ileus. Three days after admission, the patient became febrile to 102.2 F and progressively hypoxemic, requiring non-invasive positive pressure ventilation. Chest x-ray again was unremarkable. The patient became progressively hypotensive despite fluid resuscitation and hypoxemic, thus was intubated and started on intravenous antibiotics, pressor support and stress dose steroids. CT chest showed multifocal infiltrates. Blood and urine cultures grew enterococcus faecalis. Sputum cultures grew Klebsiella and Citrobacter. Echocardiogram was negative for vegetations. The patient improved clinically over the next 3 days, thus was extubated, and initiated on steroid taper. Three days after extubation, he was again febrile (101.1 F), increasingly tachycardic and hypotensive, with copious sputum production. CT chest obtained showed improvement in multifocal pneumonia but worsening patchy pneumonitis. Due to worsening respiratory and mental status, the patient was re-intubated. Sputum cultures grew Vancomycin resistant enterococci but also reported presumptive strongyloides on gram stain. The same day, larvae were also grossly visible in the patient’s urine. Ova and parasite studies on the sputum and stool were positive, with final identification of Strongyloides stercoralis larvae. Serum strongyloides IgG antibody was 3 IV (ref <0.9 IV). HIV, HTLV and quantiferon tests were negative. Patient was started on oral ivermectin and steroids were tapered due to concern of worsening disseminated disease, with gradual clinical improvement. The patient was extubated 6 days later. Patient’s stool eventually became negative for strongyloides 8 days after initiation of therapy, and he was discharged to rehabilitation nearly a month after admission.

Discussion: Strongyloides hyperinfection syndrome can result due to dissemination of larvae to multiple organs in immunocompromised hosts, with the resultant inflammation leading to systemic manifestations and even septic shock. Eosinophilia may not be pronounced in patients on steroids. Clinicians should consider testing in patients with unexplained pulmonary or gastrointestinal symptoms with epidemiologic exposure, even in the absence of eosinophilia.
Cryptococcal Meningitis in an HIV-negative patient with underlying zinc deficiency following partial gastric resection

Authors: Joseph Moran, DO; Jaimin Patel, DO; Shannon Kearney, DO

Introduction: Cryptococcal meningitis in patient’s without HIV infection is rare and most commonly occurs with a history of glucocorticoid therapy, solid organ transplantation, or hematologic cancer. In select cases, a cause of immunodeficiency is difficult to ascertain and thus, cryptococcal disease is unlikely to be suspected. This case presents a patient who was diagnosed with Cryptococcal meningitis secondary to an uncommon and often unrecognized cause of immune system compromise.

Case Presentation: An 88-year-old female with a history of partial gastric resection secondary to a strangulated paraesophageal hernia presented with generalized weakness and intermittent headache. She was admitted with similar complaints one month prior with unrevealing workup. Vital signs were normal; however, labs were remarkable for hyponatremia with a sodium level of 124 mmol/L. After admission, she became progressively encephalopathic despite appropriate correction of hyponatremia. On hospital day #8, lumbar puncture was performed and cerebrospinal fluid testing revealed the presence of Cryptococcus neoformans/gattii leading to a diagnosis of Cryptococcal Meningitis. Induction therapy with Amphotericin B and Flucytosine was initiated and serial lumbar punctures were performed to trend opening pressure. The patient had no history of HIV, took no immunosuppressive medications and had no documented history of immunodeficiency. Conversely, chart review revealed a persistent lymphopenia of unknown etiology. Her absolute CD8 and CD4 counts were profoundly low at 8 cells/mm3 and 114 cells/mm3 respectively. Immunology consultation was obtained. It was noted that the patient had normal lymphocyte counts until gastric resection 6 years prior to the current presentation. Following resection, lymphopenia persisted. Tests for vitamin and mineral deficiencies revealed zinc deficiency and replacement commenced with resolution of lymphopenia. The patient had an extended hospital course complicated by severe deconditioning, respiratory failure and hypotension and ultimately succumbed to her illness.

Discussion: Cryptococcal meningitis in patients thought to be immunocompetent presents several clinical challenges. Due to lack of clinical suspicion, diagnosis is frequently delayed which contributes to mortality rates of up to 27%, significantly higher than that of HIV patients who are diagnosed. This patient had an underlying lymphopenia following gastric resection; yet, was considered to be immunocompetent. Frequently, micronutrient deficiencies are not recognized as a cause of immunodeficiency. Zinc deficiency causes immune depression and thymic atrophy with a consequent reduction in circulating T-lymphocytes, leaving patients susceptible to opportunistic infections. This case highlights the importance of increased awareness amongst clinicians of often unrecognized causes of immunodeficiency such as zinc deficiency and the corresponding risk of opportunistic infection. Specifically, the case demonstrates the importance of close monitoring of immune function in patients with history of gastric resections. A documented history of immunodeficiency in this patient would likely have resulted in inclusion of cryptococcal disease on the initial differential, an earlier diagnosis, and a better clinical outcome.

References

Introduction: Necrotizing soft tissue infections (NSTIs) must be recognized early to prevent disability and death. The diagnosis is often delayed because of poor sensitivity of objective findings, anchoring alternate diagnoses, and lack of clinician awareness. We present an uncommon presentation of an NSTI.

Case Presentation: A 74-year-old female presented to an outside hospital with paralysis of the right upper extremity. On initial evaluation, a capillary blood glucose was >500, and further laboratory indices were consistent with diabetic ketoacidosis. Computerized tomography of the chest revealed pneumomediastinum without concomitant pneumothorax. There was extensive subcutaneous emphysema in the neck and surrounding tissues that appeared to splay from the superior mediastinum. CT imaging of the brain was negative for ischemic and hemorrhagic stroke. She was transferred to our facility for further evaluation.

On arrival to our facility, she was hypotensive and in respiratory distress. The trachea was intubated, central and arterial lines were placed, and vasopressor support was initiated. The patient was comatose with no response to central or peripheral noxious stimuli, and ecchymosis of the right neck was appreciated.

Laboratory studies revealed a severe metabolic acidosis and leukocytosis with a left shift. Blood cultures were drawn, broad spectrum antibiotics were infused, and the patient was volume resuscitated.

Diagnostic EGD and bronchoscopy were performed, and there was no evidence of macro-perforation in either the trachea or esophagus.

Over the next several hours, she remained profoundly hypotensive, and the ecchymosis had spread from her neck to the distal right upper extremity. Repeat CT scans were obtained.

Chest CT revealed predominantly anterior pneumomediastinum and perifacial fluid in the right upper arm, and CT of the abdomen demonstrated colonic wall edema thought to be secondary to ischemic injury.

Bedside arm incisions exposed grey fascia and poorly adherent soft tissue, consistent with necrotizing soft tissue infection. Exploration of the abdomen and right arm were recommended, but the family elected to forego surgery and instead focus on comfort-based care, consistent with the patient’s prior wishes. She died shortly thereafter.

Discussion: Among patients with NSTIs, the most common physical findings are pain, swelling, and fever. Soft tissue gas seen radiographically is seen in a small percentage of confirmed cases of NSTIs. The source of our patient’s subcutaneous gas was initially thought to be from transecting mediastinal air when the opposite was true. Necrotizing soft tissue infection should be considered when gas is seen within tissues or in closed compartments, especially in the presence of shock and multi-system organ failure.

Early recognition of NSTI is key to improving outcomes. Early surgical consultation should be obtained in any case of suspected NSTI. When evaluating air within tissues or any closed compartment, gas forming infections should be included in the differential diagnosis.
Acute myocardial infarction in catastrophic antiphospholipid antibody syndrome: An unusual presentation of a rare syndrome.

Authors: Meghana Parsi, MD, Maitreyee Rai, MD

Introduction: The antiphospholipid syndrome (APS) is a multisystem autoimmune condition characterized by recurrent thrombosis and/or recurrent pregnancy loss. Its clinical manifestations range from minor clots to a rapidly progressive form characterized by clots involving multiple organ systems, termed catastrophic antiphospholipid syndrome (CAPS). This occurs in 0.8–1% of all cases of APS, with a mortality rate of 50% (1). Deep vein thrombosis, pulmonary thromboembolism, and stroke are the most common manifestations. However, this syndrome rarely begins in the coronary arteries(2). We report a rare case of CAPS presenting with myocardial infarction (MI).

Case Presentation: A 69-year-old Hispanic woman with a past medical history of hypertension, diabetes, right lower extremity below knee amputation, and peripheral vascular disease requiring lower extremity bypass surgery, presented with right arm pain and swelling. She was febrile, tachycardic, and had a diffusely edematous and tender right arm. There were decreased breath sounds bilaterally, along with diminished pulses in the left foot. Labs revealed anemia, coagulopathy, acute renal failure, and elevated troponins. Chest X-ray showed lower lobe consolidation and EKG revealed a posterior wall ST elevation MI. ECHO revealed anterolateral and inferolateral hypokinesis with reduced ejection fraction. Dopplers demonstrated clots in the right basilic, cephalic, axillary and brachial veins. Antibiotics were initiated for infection and cardiac catheterization demonstrated 100% left circumflex occlusion. Attempts at cardiac stenting and revascularization, however, were unsuccessful. Surprisingly, the patient never complained of chest pain. Hospital course was complicated by hypoxia requiring intubation and gangrene of the left middle finger. Immunological studies revealed persistently elevated ANA, anticardiolipin, lupus anticoagulant and beta 2 glycoproteins. The patient was diagnosed with CAPS and improved after anticoagulation, steroids and plasmapheresis.

Discussion: CAPS is a rare variant of a well-known disease, and is likely an underreported cause of multiorgan failure. CAPS has a multifactorial etiology: 46% have primary APS, 40% have SLE, and 9% have another underlying autoimmune disorder. Often, a triggering event (infection, trauma, malignancy) is needed to incite the inflammatory process, and in our case, it may have been pneumonia. Cardiac manifestations of APS include myocardial microthrombosis, valve disease and intracardiac thrombus, with MI being reported in only 4% of individuals. Increased titers of IgG anticardiolipin antibody is thought to correlate with increased thrombosis. Recent studies even support the role of APS autoantibodies in the uptake of oxidized LDL into macrophages, resulting in enhanced atherosclerosis (3,4). While angioplasty may be considered the treatment of choice for MI, APS patients have increased risk of coronary stent thrombosis. Anticoagulant therapy started immediately after PTCA can contribute to long term coronary patency (5).

Conclusion: Given the high fatality rate of CAPS, clinicians must have a high index of suspicion for diagnosing APS in patients presenting with thrombosis of multiple organs.

References

Atypical anti-GBM disease with diffuse pulmonary hemorrhage and negative serology

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Introduction: Anti glomerular basement membrane (GBM) antibody disease is a rare disorder characterized by circulating antibodies against glomerular basement membrane resulting in rapidly progressive glomerulonephritis (RPGN) with or without pulmonary hemorrhage. Delayed or incomplete treatment can lead to dialysis dependency and death. There are few reported cases of atypical anti-GBM disease where kidneys and/or lungs are involved without evidence of circulating antibodies.

Case Presentation: A 73-year-old, non-diabetic Caucasian male with history of hypertension and stable stage G3a A1 chronic kidney disease presented for evaluation of new onset nephritic syndrome and dialysis requiring acute kidney injury (AKI). His only complaint was hematuria and mild edema without respiratory symptoms. Kidney biopsy showed non-crescentic, focal segmental membranoproliferative glomerulonephritis with a strong diffuse linear GBM staining with IgG (+3) without electron microscopy evidence of fibrils suggestive of fibrillary glomerulonephritis (GN) and lack of tubular reticular inclusions or other. He had negative serologies including a total of four anti-GBM antibodies by ELISA, antibody to the Goodpasture antigen (NC1 domain of the alpha3 chain of type IV collaged by Western Blot analysis), antineutrophil cytoplasmic antibodies (ANCA), antinuclear antibodies (ANA), C3, C4, Cryoglobulin and serum and urine electrophoresis. We proceeded with plasmapheresis and pulse steroids despite negative serologies. This decision was made in the view of dialysis dependent RPGN mimicking typical anti-GBM disease and lack of other causes of diffuse linear IgG deposition such as diabetes or fibrillary GN. Cyclophosphamide was not given as he developed E coli bacteremia from urinary tract infection. 3 weeks later, while still on 60 mg daily of Prednisone, patient presented with hemoptysis and severe respiratory distress due to diffuse pulmonary hemorrhage. He was treated with 7 sessions of plasmapheresis, pulse steroids and Cytoxan induction. He remained dialysis dependent and was discharged in stable condition to complete 9 months of Azathioprine.

Discussion: Limited data exists regarding pathophysiology, diagnosis and management of atypical anti-GBM disease. Regarding course of the disease, reported cases describe a relatively benign renal course and uncommon pulmonary involvement. This case was unique given its aggressive presentation mimicking the typical form with dialysis dependency at presentation and subsequent pulmonary involvement suggesting a common pathway and need for aggressive therapy. More studies are needed regarding potential novel assays and possible novel antibody in order to guide therapy and prognosis.
Stress Cardiomyopathy (SCM) after Electrical Cardioversion: Iatrogenic Heartbreak

Authors: Muhammad Siddique Pir, MD; Fouzia Oza, MD; Najam Saqib, MD; Madhava Rao, MD

Introduction: Stress Cardiomyopathy (SCM) is a known cause of transient left ventricular systolic dysfunction secondary to possible disruption and hyperactivation of a local sympathetic terminal with norepinephrine seethe and spillover after physical or mental insult. Direct current cardioversion (DCCV) even though a benign procedure to treat atrial fibrillation (A-fib) may, in rare cases, lead to significant left ventricular (LV) dysfunction by inducing SCM.

Case Presentation: A 81-year-old female with a history of hypertension presented to emergency department with complaints of shortness of breath for five days. She was found to have new onset atrial fibrillation with rapid ventricular response (RVR) and evidence of pulmonary edema on chest X-ray. Initial EKG showed A-fib without dynamic ST-T wave changes. Significant laboratory values included a negative high sensitivity troponin of 8ng/L with elevated Pro-BNP of 2807 pg/mL. Pharmacologic rate and rhythm control strategies were unsuccessful and patient underwent transesophageal echocardiogram (TEE) guided direct current cardioversion. TEE ruled out left atrial thrombus and showed normal left ventricular ejection fraction (LVEF). Patient was cardioverted at 150J to sinus rhythm. 48 hours after DCCV, patient exhibited clinical signs and symptoms of florid congestive heart failure. Chest X-ray showed worsening pulmonary edema and EKG showed new diffuse t-wave inversions. Pro-BNP was significant at 15,532 pg/mL. Further investigation of new EKG changes and worsening pulmonary edema led to coronary artery catheterization which showed increased left ventricular end diastolic pressure but non-significant coronary artery stenosis. Patient continued to show clinical significant signs and symptoms of heart failure. A transthoracic echo was then performed which showed LVEF of 20% with apical, anterior, inferior segment akinesis sparing the basal segment consistent with the diagnoses of stress cardiomyopathy post DCCV. Patient was treated with diuresis and guidelines directed medical therapy with improvement in her symptoms.

Discussion: DCCV for paroxysmal A-fib has been shown to cause SCM in rare cases. Pathogenesis of this phenomenon has not been defined in literature, however, few mechanisms have been proposed including disruption of local sympathetic terminals and catecholamine surge. In patients who experience worsening symptoms of heart failure after DCCV, a TTE should be done promptly to exclude SCM regardless of preserved LVEF on a prior TEE.
Immune Related Adverse Events: Isolated ACTH Deficiency with Nivolumab/Ipilimumab Combination Therapy

Authors: Joseph Scuorzo DO, Angela Magdaleno DO, Sharmila Koshy MD

Introduction: Immunotherapies have become revolutionary treatment modalities for several malignancies including patients with advanced melanoma and non-small cell lung carcinoma. New immunotherapy treatments can result in unique adverse events due to increased inflammation in any organ in the body. Common immune-related adverse events include vitiligo, colitis, hepatitis, pneumonitis, and endocrinopathies, most commonly thyroiditis.

Case Presentation: A 77-year-old male with metastatic choroidal melanoma treated with 2 cycles of Ipilimumab and Nivolumab presented to the hospital with complaints of generalized weakness, malaise, loss of appetite and 20lb weight loss over a two-week period. His concerns were initially attributed to progression of his known malignancy, however he developed further symptoms of intermittent fevers, hypotension, disorientation, tremors, and incoherent speech. Further work-up with MRI brain showed no new metastases or abnormalities and EEG was without epileptic foci. The patient gradually developed worsening lethargy, urinary incontinence, spiking fevers, and eosinophilic leukocytosis. Infectious workup was unrevealing except for a questionable right lower lobe infiltrate on chest x-ray, which was subsequently treated with antibiotics. CT chest/abdomen/pelvis showed pulmonary metastases without further abnormalities. With symptoms concerning for possible adrenal insufficiency, a random cortisol was checked resulting at 1.7 ug/dL. Adrenocorticotropic hormone (ACTH) stimulation test confirmed the diagnosis of adrenal insufficiency with the highest resultant level of 9.3 ug/dL. A low ACTH level of <5 confirmed the diagnosis of secondary adrenal insufficiency. Treatment was initiated with methylprednisolone 60mg intravenously every 12 hours with concern for autoimmune hypophysitis related to immunotherapy. Further Endocrine evaluation revealed mild TSH elevation but otherwise normal FSH, LH, Prolactin, and insulin growth factor-1. Symptoms began improving rapidly with steroid initiation.

Discussion: This case uniquely exhibits immunotherapy induced isolated ACTH deficiency, rather than hypophysitis. Hypophysitis can occur with an adverse event rate up to 17% while on Ipilimumab, however isolated ACTH deficiency is rarer. Diagnosis is challenging with symptomatology mimicking common malignancy complaints. It is important for all physicians to be able to recognize symptoms of secondary adrenal insufficiency, especially as a side effect of immunotherapy, as this condition is life threatening and requires immediate diagnosis and treatment.
Adult-onset Still’s disease with Macrophage Activation Syndrome

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Introduction: Adult-onset Still’s disease (ASD) is an inflammatory disorder characterized by arthritis, daily fevers, and evanescent rash. Age of onset is over the age of 16. ASD can be associated with the Macrophage activation syndrome (MAS), which can be a fatal syndrome of excessive immune activation. It is Hemophagocytic lymphohistiocytosis in the setting of rheumatic disease and should be treated as a life-threatening emergency.

Case Presentation: A 52-year-old, who had a history of inflammatory arthritis being treated with methotrexate and prednisone, presented to ED for a six-month history of nightly fevers of 38.3 degrees Celsius, chills, night sweats, and general malaise. He progressively worsened with pleuritic chest pain associated with dry cough and shortness of breath. He was admitted and initiated on antibiotic therapy for suspected sepsis secondary to pneumonia. During the admission, the patient developed new hoarse voice and pain in his toes and knees. Infectious work up, including TB, HIV, and Malaria were negative. Serum ferritin was found to be 18,581ng/mL. His fevers waxed and waned. They continued at the time of discharge. Arthralgias in his toes had improved but never resolved.

He returned to the ER five months later with progressively worsening night sweats, chills, weight loss, body aches, lymphadenopathy, and weakness. He developed fever with rigor. His blood work revealed Hemoglobin of 5.4 g/dL, creatinine of 1.2 mg/dL, lactic acid level of 2.6 mmol/L, as well as significant leukocytosis of >68 k/uL, with 93% neutrophils with left shift. LFTs showed AST of 55 U/L, ALT of 17 U/L, and ALP of 134 U/L. His CXR showed bilateral opacities. He, subsequently, developed respiratory failure, requiring intubation and was transferred to ICU. It was observed that he had waxing and waning of salmon-colored rash that coincides with occurrence of fever. In the setting of arthralgia, spiking fevers, and evanescent rash, a diagnosis of Adult onset Still’s disease was made. Peripheral smear, then, showed phagocytes of neutrophils. Repeated serum ferritin was >40,000ng/mL. He was then diagnosed with macrophage activation syndrome. Due to being critically ill, he was transferred to a tertiary care center for further care. There, he was found to have significant EBV viremia via PCR. Unfortunately, the patient expired two days after transfer.

Discussion: This case illustrates the difficulty in diagnosing MAS due to its infrequency. The length of time to diagnosis is often protracted secondary to lack of physician awareness. Moreover, the nature of adult onset still’s disease is primarily a diagnosis of exclusion. Improved recognition of the combination of symptoms can help to minimize morbidity and mortality in patients with this disease.

References

Spontaneous Hemorrhage in a Young Woman with Chronic Myelogenous Leukemia (CML)

Authors: JC Shih, MD; ME Bromberg, MD

Introduction: CML is a clonal myeloproliferative neoplasm associated with the formation of the BCR-ABL fusion gene. Although not often appreciated, bleeding is a common clinical feature of CML and may be due to platelet dysfunction.

Case Presentation: A 25-year-old woman with Philadelphia chromosome positive CML presented with progressive onset of left lower extremity pain and swelling over one week which eventually prevented her from ambulating. Of note, one month prior, she was admitted to an outside hospital for a leukocyte count of 488x10^9/L with questionable compliance to imatinib and was discharged after her CML was controlled with imatinib and hydroxyurea.

On exam, she was notably afebrile and non-toxic appearing. There was significant swelling, erythema and tenderness of the left lower extremity below the knee. Her spleen tip was palpable 12 cm below the left costal margin. A CBC showed a leukocyte count of 371x10^9/L, hemoglobin of 7.1 g/dL, and platelet count of 186x10^9/L. Her PT and aPTT were normal. Imaging of her left lower extremity revealed multiple complex fluid collections in the left proximal mid-calf musculature. Aspiration of one of the fluid collections demonstrated only minimal sanguineous fluid. Due to persistent swelling and pain, she was taken to the operating room (OR) for exploration which revealed a hematoma. She was taken back to the OR two days later due to an acute decline in hemoglobin level with signs of compartment syndrome and underwent a fasciotomy. Post-operatively, she developed an additional hematoma in her left upper extremity and continued bleeding from her lower extremity surgical wound requiring multiple transfusions despite normal platelet count, PT, aPTT, factor VIII activity level, von Willebrand’s antigen level and ristocetin cofactor activity. Due to the possibility of a qualitative platelet defect, she received multiple platelet transfusions which markedly reduced her bleeding.

Discussion: In a report of clinical features at diagnosis of 430 patients with CML, bleeding (21.3%) was only second to fatigue (33.5%) (1). Moreover, formation of the BCR-ABL fusion gene may result in a clonal population of megakaryocytes that produce dysfunctional platelets (2). Also, some tyrosine kinase inhibitors (TKIs) used to treat CML can cause platelet dysfunction (3). Results of platelet aggregation testing in 91 patients with CML demonstrated impaired platelet aggregation in 85% of patients on dasatinib and 67% of patients on imatinib (4).

Our patient with no prior history of bleeding abnormalities, presented with spontaneous hematomas in the setting of normal platelet count, coagulation studies and von Willebrand’s panel. She demonstrated improvement of her bleeding after aggressive platelet transfusions suggesting a qualitative platelet defect. This case highlights that despite normal platelet counts, bleeding can occur due to acquired or therapy-induced platelet dysfunction in CML and that platelet transfusions are an important maneuver to improve outcomes.

References

Esophageal Necrosis: A Rare Complication of Diabetic Ketoacidosis – A Case Report

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Introduction: Diabetic ketoacidosis (DKA) is a common complication of uncontrolled diabetes mellitus. Rarely, DKA can be complicated by acute esophageal necrosis (AEN) with the hallmark finding of circumferential black distal esophageal mucosa seen with endoscopy. Common presentation of AEN includes upper GI bleeding, nausea, vomiting, dysphagia, and syncope. Most commonly AEN is associated with hemodynamic compromise and low flow states, like what is encountered with the profound dehydration in patients with DKA. Those considered at greatest risk for developing AEN are men over age 50 with comorbid conditions such as DM, malignancy, hypertension, alcohol abuse, and CAD.

Case Presentation: A 50 year old male with past medical history of DM type I, gastroparesis, chronic pancreatitis, and Barrett’s esophagus presented with non-radiating epigastric abdominal pain, coffee ground emesis, and nausea. He was vomiting frequently with severe pain for approximately two days. He denied any new medications, sick contacts, alcohol intake or intentional ingestions at the time. Arriving vitals were systolic BP 60’s – 70’s, HR 98, GCS 12. Dried blood was present around the patient’s mouth and tenderness in the epigastric area was present. He was given IV normal saline for volume resuscitation as well as emergent blood transfusions. He was started on empiric treatment for a GI bleed along with Zofran for persistent nausea. Initial lab work revealed hemoglobin 9.9, blood pH 7.34, lactate 3.7, glucose >1000, positive serum ketones, bicarbonate 14, and anion gap 39. CT scan of his abdomen and pelvis revealed esophageal wall thickening, gastric distention with dilated fluid-filled esophagus, fatty liver, and chronic pancreatitis. He was later admitted to the MICU due to his critical clinical status.

A diagnosis of diabetic ketoacidosis was made and DKA treatment protocol with insulin infusion, electrolyte replacement and frequent labs was started. Gastroenterology consult with EGD revealed the distal 2/3 of the esophagus having black, necrotic appearing mucosa with abrupt transition at the GE junction. No biopsies were taken due to the nature of the patient’s presentation and risk of perforation. The patient was subsequently started on IV PPI and Carafate with gradual improvement of his symptoms.

Discussion: AEN is a rare complication of DKA and was discovered in this patient. Typical management involves fluid resuscitation, treatment of the underlying DKA, PPI, Carafate, and keeping the patient NPO. Although uncommon, AEN is an important condition to recognize as it is associated with mortality rates ranging from 15% - 36%. Also, 15% of survivors develop long term complications such as esophageal strictures or stenosis. Many of the symptoms of AEN are common presenting symptoms of DKA so clinical suspicion and evaluation in the correct context could prevent morbidity and mortality.

References

Pylephlebitis, gram negative sepsis and cyclical wheezing- A case for Strongyloidiasis.

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Introduction: Strongyloides stercoralis is a parasitic nematode which is associated closely with gram negative bacteremia particularly in the setting of hyperinfection. The dissemination of the larva via the bloodstream and to various end organs causes an inflammatory response which manifests clinically as hyperinfection syndrome. Portal venous gas and pylephlebitis are seen in complicated intra-abdominal sepsis. We present the case of pylephlebitis with E.coli bacteremia in the setting of underlying strongyloidiasis.

Case Presentation: A 59-year-old gentleman presented with acute onset fever, chills, abdominal distension and cholestatic jaundice. He was previously healthy, had no sick contacts and no recent travel. At presentation he met SIRS criteria for sepsis and our initial differentials included cholecystitis, cholangitis, and choledochohilitis. However this patient denied having any abdominal pain. There was no history of gallstones, prior surgeries. Labs were significant for leucocytosis, direct hyperbilirubinemia and mild AST/ALT elevation but no ALP elevation. Imaging ruled out obstruction in the biliary tract and cholecystitis. Instead this patient had hepatic portal venous gas, pylephlebitis and diverticulitis. Presence of gas in the portal venous tract is associated with high morbidity and mortality. Pylephlebitis is a suppurative infection of the portal vein which is associated with intra-abdominal infections, and is related to bacteremia most commonly with Bacteroides or E.coli. Our patient had E.coli bacteremia and was treated with intravenous antibiotics. During the hospitalization he was noted to have short, intermittent, self-limited episodes of wheezing and rigors which he later told us had been occurring infrequently for several weeks. Cyclical wheezing in the setting of gram-negative sepsis prompted us to consider the possibility of a parasitic infection such as Strongyloides. Subsequently on probing his travel history we found that he had traveled extensively in the remote past over 30 years back. Serological testing for Strongyloides came back positive for the patient.

Discussion: We present a case of E.coli bacteremia and pylephlebitis. which was initially attributed to diverticulitis with micro- perforation and ascending infection. However, the patient developed episodic wheezing and rigors. This finding in a patient originally from a hyperendemic strongyloidiasis area is better elucidated by an underlying Strongyloidiasis hyperinfection syndrome. Strongyloidiasis is a possibility even if the exposure was many decades ago as evidenced in our patient. Additionally, this patient had pylephlebitis which is a rare association with Strongyloides and has only been documented in a few case reports. This case report illustrates the clinical setting in which to suspect a Strongyloides infection and includes a discussion about the diagnosis and management of the same.

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Urinoma with Unilateral Urinothorax: A Very Rare Cause of Abdominal Pain and Pleural Effusion

Authors: Anika Toor MD, Nihit Shah MD, Bindiya Shah MD, Amit Toor MD, Mahesh Krishnamurthy MD

Introduction: Urinoma is the result of retroperitoneal leakage of urine from a disruption of the urinary tract. Urinomas can be obstructive or traumatic. The most commonly accepted mechanism is the backflow of urine with increased intra-pelvic pressures and subsequent fornixal rupture. On the contrary, it is believed to have a renoprotective effect in the long-term. We present an extremely rare case of abdominal pain and pleural effusion occurring from urinoma with a resultant unilateral urinothorax.

Case Presentation: The patient is a 76-year-old male who presented with generalized weakness, anorexia and abdominal pain. Medical history is pertinent for prostate cancer 20 years ago status post radiotherapy. On physical exam, he appeared frail and dehydrated. Labs revealed anemia and renal failure with a serum creatinine of 8.7 mg/dL and potassium of 8.6 mmol/L. He required hemodialysis for acute renal failure and severe hyperkalemia. Work up including complement levels, anti-neutrophil cytoplasmic antibody, anti-nuclear antibody, anti-myeloperoxidase antibody, and anti-proteinase 3 antibody was negative. CT abdomen revealed right pleural effusion and a large 25 cm size retroperitoneal fluid collection posterior to the right kidney, suspicious for urinoma. Thoracentesis revealed transudative effusion with a low pH. A pleural fluid creatinine to serum creatinine ratio was 1.07, which is consistent with the diagnosis of urinothorax. The patient underwent retroperitoneal fluid drainage and nephroureterostomy tube placement. Elevated retroperitoneal fluid creatinine of 36.6 mg/dL to serum creatinine of 5.1 mg/dL was suggestive of urinoma. A renal nuclear scan with MAG-3 revealed a leak of the radiotracer through the right kidney and into the retroperitoneal fluid drainage bag, confirming this complicated diagnosis of urinoma. His serum creatinine trended down to 1.0 mg/dL with relief of urinary obstruction. Our patient is currently off of hemodialysis. He underwent cystoscopy at a later date showing a high-grade transitional cell carcinoma of the bladder.

Discussion: Urinomas can be asymptomatic or present with non-specific symptoms such as abdominal pain, malaise, and oliguria. A high index of clinical suspicion is important for early diagnosis and optimal management. Transudative pleural effusion with a low pH is one of the leading clues to this diagnosis. Urinothorax should be suspected in the presence of unexplained pleural effusion and obstructive uropathy. CT abdomen with contrast is the best modality for diagnosis. Small urinomas are managed conservatively, whereas large and obstructive urinomas require drainage through a urological procedure.
Successful Treatment of HIV-Associated Pulmonary Arterial Hypertension After Initially Misdiagnosed by Right Heart Catheterization

Authors: Cyrus Vahdatpour, Timothy Barry, Christina Malouf, and Edward Wu

Introduction: Pulmonary Hypertension (PH) in patients with HIV is commonly a result of either left-sided heart disease or HIV associated Pulmonary Arterial Hypertension (PAH). PH due to left-sided heart disease reflects the aging HIV population due to advances in medical care; and is managed by addressing the underlying pathology from the left side of the heart. Approximately 0.5% of the HIV population develops PAH and there are multiple pharmacological options specific to PAH (1). Most patients with HIV-associated PAH are diagnosed late in their disease course and, as a result, the natural history of this disease remains unknown. We present a diagnostically challenging case of HIV-associated PAH, initially diagnosed as PH secondary to left-sided heart disease.

Case Presentation: A 44-year old HIV-positive male presented to the emergency department with a several month history of progressive dyspnea on exertion. Physical examination was significant for a heart rate of 112 beats per minute, elevated jugular venous pressure, and ascites, consistent with gross volume overload. Laboratory data revealed an elevated NT-proBNP of 1540 pg/mL, and an elevated creatinine of 1.32 mg/dL, signifying acute kidney injury. An EKG showed sinus tachycardia, right bundle branch block, and right axis deviation. Chest radiography demonstrated a right-sided pleural effusion. Transthoracic echocardiography demonstrated moderately elevated pulmonary artery pressures, an enlarged right atrium and ventricle with a displaced septum, severe tricuspid regurgitation, and reduced left ventricular ejection fraction. Right heart catheterization (RHC) revealed elevated left- and right-sided pressures (mean PA pressure of 46, wedge of 20, diastolic pulmonary gradient of 18, PVR of 10 woods units, and cardiac index of 1.28). After optimization of the patient’s volume status with diuretics, dobutamine, sildenafil, and hydralazine, the indwelling RHC revealed pressures consistent with group 1 PAH (mean PA pressure of 29, wedge of 8, diastolic pulmonary gradient of 15, PVR of 6.4, and cardiac index of 1.7). The patient was discharged after two weeks of titration of sildenafil and weaning off of hydralazine, dobutamine, and diuretics.

As an outpatient, ambrisentan was added to sildenafil and the dosage was increased until he had an improved 6-minute walk distance of approximately 100 meters from baseline and improvement from World Health Organization Class IV to II.

Discussion: PAH is often diagnosed late in its disease course due to its insidious onset. Echocardiography may suggest left-sided heart etiology in patients with severe volume overload, and ventricular interdependence may be difficult to interpret based on limited clinical context. These results often affect cardiac catheterization interpretation, requiring increased clinical suspicion and fluid optimization, followed by repeat catheterization. While RHC is debated in regards to safety outweighing clinical impact, it has been associated with reduced mortality when it impacts medical management (2).

References

Pemrolizumab triggered Myasthenic crisis: A near fatal respiratory failure

Authors: Selamawit Woldemariam, MD, Mahesh Krishnamurthy, MD, Shumona Ima, MD, Divakar Sharma, MD

Introduction: Metastatic melanoma was historically considered a uniformly and rapidly lethal malignancy with limited options for treatment until the introduction of checkpoint inhibitors as immunotherapy in recent years. These agents have been largely tolerated and have the potential to result in durable responses however it is important to recognize the unique side effect profiles that lead to triggering or exacerbation of underlying autoimmune diseases. Pemrolizumab is a PD 1 inhibitor that was approved by the FDA in 2014 being the second checkpoint inhibitor approved in the United States.

Case Presentation: We present the case of an 81-year-old male patient who presented for an episode of aspiration and altered mental status. History suggested progressive generalized muscle weakness and fatigue associated with difficulty swallowing which manifested a few days after initiation of therapy for melanoma with Pemrolizumab. The patient had prior history of Seropositive myasthenia gravis but was in remission and had stopped treatment for more than a year. Records revealed history of BRAF negative metastatic melanoma of left forearm with liver and brain metastases diagnosed 8 months ago and pemrolizumab initiation 10 weeks ago. A few hours in to hospitalization and initiation of therapy for aspiration pneumonia, patient developed sudden respiratory failure requiring mechanical ventilation and was transferred to ICU. He was started on therapy for myasthenic crisis and did show significant daily improvement. Negative Inspiratory force measurements were monitored to assess respiratory muscle capacity. Although significant clinical improvement was made, considering patient’s age, multiple comorbidities and poor prognosis, decision was later made to transition him to hospice care and patient had expired shortly after this.

Discussion: More and more cancer therapies are emerging targeting the immune system rather than the cancer cells themselves. One of these are antibodies that block specific immune checkpoint molecules of programmed cell death protein (PD1) and its ligand PD-L1. Although satisfactory antitumor results have been documented with these medications, in addition to Dysimmune toxicities called immune related adverse events (irAE), these medications also trigger or exacerbate autoimmune diseases. Careful assessment for underlying autoimmune diseases should be done prior to initiation of treatment.
The Hemolysis challenge - Post Babesiosis Autoimmune hemolytic anemia in a patient with sickle cell disease

Authors: Tanuja Yalamarti, MD, Yii Chun Khiew, MD, Melinda Darnell, MD

Introduction: We present a diagnostically challenging case of a sickle cell patient who acquired Babesia infection due to blood transfusion but acquired secondary hemolytic anemia after resolution of the infection.

Case Presentation: 27 year-old female with a history of sickle cell anemia (type SS) on bimonthly apheresis, pulmonary hypertension, and prior stroke was admitted with a fever (Tmax 102F) and lower extremity pain symptoms suggestive of vaso occlusive crisis.

A peripheral blood smear incidentally detected parasites and further confirmatory tests revealed intraerythrocytic forms consistent with Babesia with a parasitemia level of 1.6%. Malaria antigen test was negative. As the patient denied any prior travel history, the parasitemia was hypothesized to have been acquired through prior blood transfusions. She completed a full course of Atovaquone and Azithromycin. Two weeks after the diagnosis of babesiosis, her hemoglobin dropped from 8.4 to 5.4 mg/dl, and repeat peripheral smears confirmed clearance of parasitemia. Other laboratory tests included elevated lactate dehydrogenase level of 750U per liter, a reticulocyte count of 28%, and undetectable haptoglobin consistent with hemolysis. Blood-bank testing was notable for a positive direct antiglobulin test for IgG and complement component 3 (C3). The underlying cause for recrudescent hemolysis in the setting of parasitemia clearance was thought to be due to autoimmune hemolysis. Further studies did not detect new alloantibodies. Warm autoantibodies were identified.

Given the confounding factors of chronic hemolysis and frequent transfusion requirements in the setting of sickle cell anemia, the contribution of warm autoantibodies to her underlying acute clinical hemolysis could not be adequately assessed. Hence, the patient did not receive immunosuppressive therapy. The patient had a prolonged hospitalization, and subsequently required additional red cell exchange transfusions in order to adequately manage her vaso-occlusive crisis.

Discussion: Red blood cell transfusions have reduced the morbidity and mortality in patients with sickle cell disease, while also exposing them to numerous well-known risks. Though the prevalence of transfusion-acquired infections have significantly decreased, the risks are not completely eliminated. Babesia infection causes hemolysis by directly attacking the erythrocytes. There are limited cases in the current literature reporting warm autoimmune hemolytic anemia (WAHA) related to Babesia infection. In particular, asplenic patients are at a higher risk as they cannot efficiently remove immune complexes formed by a type III hypersensitivity reaction, thus triggering the classical complement cascade and subsequent hemolysis.

To our knowledge, this is the first case of a patient with sickle cell disease who developed WAHA due to Babesiosis and did not require immunosuppression and was discharged with an overall good outcome despite a prolonged disease course. Testing for immune-mediated hemolysis such as WAHA should be considered in patients with worsening or persistent hemolytic anemia after successful treatment of babesiosis, particularly in asplenic patients.
Metformin-Associated Lactic Acidosis precipitated by NSAID-Induced Acute Kidney Injury

Authors: Edgar Zamora MD; Tarek Hassouna MD; Silpa Kosuri MD

Introduction: Metformin is an oral biguanide antidiabetic medication used for treating type 2 diabetes mellitus. It is contraindicated in patients with renal or hepatic insufficiency due to the risk of lactic acidosis.

In the diabetic population, the incidence of lactic acidosis as a side effect of metformin is rare, with an incidence rate of 9 cases per 100,000 person-years. Most of these cases are from patients who have abnormal renal function, and this side effect is considered exceedingly rare in patients with normal renal function.

Non-steroidal anti-inflammatory drugs (NSAIDS) may be associated with renal side effects in some populations, including those with preexisting renal disease or decreased renal perfusion.

We present the case of a patient with initial normal renal function who had reportedly been taking metformin for many years and who was just recently prescribed NSAIDS believed to have caused acute kidney injury and triggered metformin-associated lactic-acidosis.

Case Presentation: An 82-year-old female with type 2 diabetes mellitus taking metformin 1000 mg twice a day and lisinopril 20 mg daily, presented to our institution complaining of nausea and vomits for the previous 4-5 days.

The patient was lethargic and short of breath. Her blood pressure was 75/58 mmHg, pH of 6.7, serum bicarbonate of 7 mEq/L, pCO2 of 21 mmHg, anion gap 37.7, creatinine was 7.7 mg/dL, hemoglobin 14.7 g/dL and lactic acid 5.8 mmol/L. Despite fluid resuscitation, she became progressively hypotensive and developed acute hypoxic respiratory failure, requiring orotracheal intubation, vasopressor medications, and admission to the intensive care unit.

Additional history revealed a recent motor vehicle accident 13 days prior to consultation, when she consulted another institution and was prescribed ibuprofen 800 mg every 6 hours, which she was taking at least 3 times a day.

Lactic acid peaked to 20 mmol/L. Treatment provided with intravenous bicarbonate, fluids, and continuous renal replacement therapy. Hemodynamic status steadily improved, creatinine and lactic acid slowly normalized by the 4th day after stopping her home medications.

Discussion: For type 2 diabetic patients, the use of metformin results in improved survival in addition to renal and cardiovascular protection. Although the risk of lactic acidosis is well known, it is rare and the use of this medication regarded as beneficial for most patients with type 2 diabetes mellitus, it should however be revised according to a GFR cut-off or adjusted determined on its plasma level concentrations.

In patients with normal renal function, NSAIDS have shown no influence in GFR or renal blood flow (RBF). Conversely, for patients with underlying diabetic nephropathy, a single dose of ibuprofen may reduce GFR and RBF. For patients with a long history of diabetes mellitus type 2, and current treatment with an angiotensin-converting enzyme inhibitor caution should be exerted on prescribing high dose NSAIDS.

References
Unusual case of Bacterial Endocarditis in a Patient without Intravenous Drug Use

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Introduction: Infective endocarditis (IE), a life-threatening condition carrying a mortality rate of 30%, seldom presents as right-sided infective endocarditis (RSIE). Such cases are overwhelmingly associated with intravenous drug use (IVDU). IE can occur on any surface of the heart's lining and vegetation is monitored via echocardiography. Below, we discuss an interesting case of RSIE with a high-risk follow-up echocardiograph in a non-IVDU.

Case Presentation: A 62 year-old female with a past medical history of diverticulitis presented with 5 days of fever, chills, and lumbago. On arrival, the patient was noted to be tachycardic (142), tachypneic (35), and febrile (103.2°F) with a blood pressure of 95/47 mmHg. Further examination revealed dry mucous membranes and negative Psoas sign. She denied IVDU. Labs showed pancytopenia with White Blood Cells 2.4 Thou/µL, Red Blood Cells 4.10 Mill/µL, Platelets of 29 Thou/µL. She also had an elevated lactate (3.9 mmol/L), total bilirubin (1.4 mg/dL). Direct bilirubin was 0.5 mg/dL with normal coagulation studies. Urine drug screen was negative.

The patient was admitted to the Intensive Care Unit for septic shock where she was treated with broad-spectrum antibiotics and fluid resuscitation. Blood cultures grew Cefazolin-sensitive Beta Hemolytic Streptococcus B. While a transesophageal echocardiogram (TEE) was indicated, the patient refused this test. A transthoracic echocardiogram (TTE) did not reveal valvular vegetation. An MR Lumbar Spine, obtained for lumbago, demonstrated early septic arthritic changes in the posterior left facets of L3 and L4. She was discharged on a 6-week course of Ceftriaxone. While in-house repeat blood cultures remained negative, the patient was readmitted the following week with similar symptomatology. CT Chest revealed right upper, middle, and lower lobe pulmonary emboli with right heart strain, which prompted a TEE. A 1.7 x 1.2 cm vegetation on the mid-basal posterior leaflet of the tricuspid valve was identified and its presence was confirmed on Cardiac MRI. Antibiotics were optimized to ampicillin and gentamicin, as per infectious disease recommendations. Following 6 weeks of antimicrobial therapy, a repeat TEE performed demonstrated an unchanged vegetation.

Discussion: RSIE, accounting for 5-10% of all IE, is strongly associated with IVDU in addition to defibrillator/pacemaker leads and dialysis catheters, none of which existed in our patient. Moreover, our patient's cultures grew Streptococcus, which is a rare cause of RSIE (50-80% of cases are Staphylococcus aureus). Follow-up of vegetation are usually performed by TTE or TEE. Although frequency and timing of these studies are based on the type of microorganism, initial findings, and response to therapy, they are commonly performed after 7 days and upon completion of the antibiotic course. It is important to remind ourselves that close monitoring of vegetation size is vital in IE patients, as an increase in size or failure to regress, despite antibiotic therapy, is a strong predictor of mortality and embolic events, as evidenced by this case.
A Missed Case of Pheochromocytoma Leading to Cholinergic Crisis and Rare Cerebrovascular Complication

Authors: Maryam Zia, MD, Salik Aleem, MD, Munira S. Abbasi, MD

Introduction: Pheochromocytoma is a rare catecholamine-secreting neuroendocrine tumor with a heterogeneous presentation. We present a case of pheochromocytoma crisis leading to multiorgan failure and stroke.

Case Presentation: A 41-year-old female with a history of Hashimoto thyroiditis, hypertension, and generalized anxiety disorder presented to an outside hospital ED with a severe frontal headache, palpitations, and nausea. CT head obtained was unremarkable. Suspecting it to be a migraine, she was given an infusion of diphenhydramine, ketorolac, and metoclopramide. Immediately after the infusion she experienced a tonic-clonic seizure and became tachycardic. She received Ativan and was intubated for airway protection. Post-ictal vital signs displayed persistent tachycardia. CTA chest pursued to rule out acute PE was negative but incidentally revealed a large right adrenal mass. Patient was transferred to a tertiary care ICU. She was started on α and subsequently β adrenoreceptor blockade. Elevated plasma metanephrines confirmed the diagnosis of pheochromocytoma. Hospital course was complicated by AKI, transaminitis, and transient cardiomyopathy in the setting of cholinergic crisis. Post-extubation she was found to have dysarthria and visual field defects. MRI brain showed cerebellar swelling and multiple bilateral subacute cerebellar, occipital, and parietal infarcts. MRA was concerning for basilar artery narrowing. CTA head was not pursued due to continuing renal injury. Interval CT head showed stable cerebellar swelling without new infarcts. The patient underwent uncomplicated right adrenalectomy. At follow up 2 months later, MRI brain showed old bilateral cerebellar and left occipital infarcts while MRA was negative for basilar artery narrowing.

Discussion: Hypertension with paroxysmal headaches, palpitations, and diaphoresis are classical symptoms of pheochromocytoma. Given the rarity of the disease and lack of awareness by providers, symptoms may be mistaken for common diagnoses such as migraine headaches, arrhythmias, or GAD. We suspect our patient presented during a symptomatic episode, and metoclopramide triggered a catecholamine surge. Multiorgan failure can be explained by acute hemodynamic disturbances. Although ischemic stroke is not well-described in the literature, there are two reported mechanisms behind cerebrovascular symptoms in pheochromocytoma. First, acute elevation in BP can lead to reversible posterior leukoencephalopathy syndrome characterized by symmetrical white matter edema in the posterior cerebral hemispheres. Second, increased catecholamine levels can cause vasospasms leading to intracerebral ischemia and infarct. Both mechanisms likely played a role in our patient as she had cerebellar edema in the occipitoparietal lobes as well as basilar artery narrowing.

Conclusion: Pheochromocytoma should be considered in the evaluation of an intermittent headache, palpitations, and hypertension. An early consideration is necessary to avoid anchor bias. Adrenergic crisis can affect nearly all organs including the brain. The source of ischemic stroke in these patients appears to be hypertensive and vasospastic related.
Importance of Adequate Screening before Immunosuppression: Leprosy Exacerbation after Treatment with Adalimumab

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Introduction: Increased use of both biologic and conventional disease-modifying antirheumatic drugs (DMARDs) has significantly improved outcomes in the treatment of inflammatory conditions; however, their use still carries the potential for serious adverse reactions, some of which may be extremely rare, making their diagnosis challenging.

Case Presentation: This is the case of a 57-year-old man without known systemic illnesses, who worked as a dishwasher, traveled twice to the Dominican Republic, lives with his wife who is Dominican, and reports chronic use of marijuana. For the past 15 years he experienced bilateral hand numbness. Ten years ago, he developed finger contractures, hand burns, and recurrent skin bullae, reason why he was diagnosed with contact dermatitis vs bullous disease. As finger contractures worsened, 7 years ago he was diagnosed with rheumatoid arthritis and started on methotrexate and adalimumab. Subsequently, 3 years later hand numbness progressed to complete loss of sensation, which he recognized after burning while cooking without feeling any pain. These burns and new erythematous patches lead to an evaluation by a Dermatologist who discontinued both methotrexate and adalimumab and performed a skin biopsy. Physical examination was remarkable for bilateral madarosis, right lagophthalmos, palpable great auricular nerves, nonreducible finger contractures, hand muscle atrophy, and infiltrative scaly erythematous plaques involving earlobes, areolas, and extensor surface of both upper and lower extremities. These erythematous plaques and hypoesthesia, in addition to a Fite stain skin biopsy showing acid fast positive bacilli, indicated the diagnosis of Hansen’s disease, mid-borderline variant. Treatment was initiated with dapsone, rifampin, clofazimine, prednisone, and methotrexate resulting in a significant improvement of skin lesions and lagophthalmos.

Discussion: Both rheumatologic and dermatologic manifestations associated with Hansen’s disease could lead to misdiagnosis. More notably, Biologics and conventional DMARDs carry the potential for serious adverse reactions such as exacerbation of a dormant disease like leprosy, which is extremely rare, but endemic in Puerto Rico. This case proves us the importance of an adequate screening that includes a detailed clinical history, complete physical exam, and relevant laboratory work up before starting immunosuppressive therapy. Screening, not only for infections such as latent tuberculosis and hepatitis B & C, but also for comorbidities like heart failure and malignancy, could allow the patient to obtain maximum benefit from immunomodulators with minimal complications. This is of importance as these drugs, particularly Biologics, have revolutionized the current treatment of inflammatory conditions, are being increasingly used in this era, and may be initiated by primary physicians without the appropriate screening.

References


Wolinsky: A rare and overlooked cause of peritoneal infection.

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Introduction: Peritonitis is one of the feared complications in patients with end-stage renal disease (ESRD), undergoing peritoneal dialysis, increasing the morbidity and mortality of these patients. The most common organisms involved are Gram positive organisms, for which are usually treated with vancomycin and an aminoglycoside.

Case Presentation: A case of an 82-year-old female patient with past medical history significant for ESRD on peritoneal dialysis was referred to the hospital from nephrologist’s office due to severe generalized abdominal pain. Patient’s symptoms had progressed for the past few weeks, and an abdominal paracentesis with peritoneal fluid analysis was performed, outpatient. After this, the patient was treated with an unknown antibiotic due to positive results without resolution of symptomatology. Upon arrival to the ER, the patient was chronically ill, in no acute distress, alert, and completely oriented. Physical examination was significant for mild abdominal distention, hyperactive bowel sounds, and diffuse tenderness upon palpation. Laboratory workup was significant for elevated creatinine level consistent with the patient’s baseline, normocytic normochromic anemia, and negative HIV test. Abdominal CT scan was performed which showed moderate mural thickening of the cecum ascending colon suggestive of inflammatory or infectious colitis, a peritoneal dialysis catheter with the distal end coiled within the left side of the pelvis, and ascites. Diagnostic paracentesis with peritoneal fluid analysis was performed, and broad-spectrum antibiotics were initiated comprising: vancomycin, piperacillin-tazobactam, and metronidazole. The peritoneal fluid analysis was significant for 309 WBC with increased polymorphonuclear cells, and culture and sensitivity analysis reported acid-fast bacilli (Mycobacterium wolinskyi) growth. Anti-infective therapy was re-adjusted to Rifampin, Isoniazid, Pyrimethamine, and Ethambutol. WBC count of intraperitoneal fluid continue to increase despite quadruple therapy, and Ciprofloxacin was then added to treatment. Patient then gradually responded to therapy with continual decrease in WBC count. Peritoneal catheter was removed, and patient was discharged on Ciprofloxacin to complete a total of 6 months outpatient.

Discussion: Mycobacterium wolinskyi has been previously described as the causative agent for post-traumatic infected injuries, potentially leading to osteomyelitis. With only 12 cases reported on this organism, and none of them related to peritoneal dialysis catheter infection, makes this presentation extremely rare in nature. Other Nontuberculous mycobacteria species have been noted to cause these types of infections, but the prevalence is yet still considered to be low. The prompt recognition of such organisms in the peritoneal fluid leads to rapid treatment and decreased morbidity and mortality.

This case is significant not only due to its rarity, but also, is a possible etiology that should be considered in a patient with recurrent peritoneal dialysis-related peritonitis end-stage prompt proper diagnosis and re-direct medical management if needed to avoid serious complications including death.
Candida duobushaemulonii: A rare but emergent cause of candidemia with a high resistance antifungal profile

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Introduction: Candida duobushaemulonii is a rare but emerging fungal pathogen. It is part of the Candida hameulonii complex and related to the highly pathogenic Candida auris. Their multiresistant antifungal profile is a concern, limiting the selection of effective therapy. In this case we report a case of a candidemia with this specie in a critically ill patient, its resistance profile and the patient’s outcome.

Case Presentation: A 71-year-old male was admitted to the intensive care unit with a large bowel perforation and sepsis after an elective colonoscopy for which he was started on ciprofloxacin and metronidazole. The patient was taken into surgery for diverting colostomy and a bogota bag was placed. After the surgery he remained in mechanical ventilation. On day #8 of hospitalization, the patient develops fever, tachycardia, and low blood pressure. Physical examination was pertinent for the presence of a femoral central line, unremarkable lung examination and an abdomen without any abnormality from baseline. Laboratories showed leukocytosis with bandemia, and absence of pyuria or bacteria in the urinalysis. The chest film had no infiltrate. Blood cultures were drawn, and he was switched to vancomycin and meropenem. Three days later, yeasts were reported in blood cultures and he was started on caspofungin (50 mg daily). After six days of cultured blood a Candida duobushaemulonii was identified via VITEK2(8.01), showing resistance to all azoles, to most echinocandins and to amphotericin B as identified by Sensititre-YeastOne. The specie was only susceptible to flucytosine (an oral therapy that cannot be given as monotherapy) and to micafungin (which was not available). Therefore, he was started on high doses of caspofungin (150 mg daily) and flucytosine, while the central line was removed. Because Candida auris can be misidentified as Candida duobushaemulonii by commercial methods a sample of the specimen was sent to CDC in Atlanta for identification. The specimen was also identified as Candida duobushaemulonii via MALDI-TOF method (Bruker Biotyper with MicrobeNet and Bruker Daltonics databases). Further blood cultures were negative after three days of therapy. Once micafungin was obtained the therapy was modified and he completed a total of eleven days of initial combination therapy and five additional days of micafungin. He tolerated the therapy well and was successfully extubated. He survived 30-day post-fungemia.

Discussion: Traditionally, Candida glabrata and Candida krusei are known for being the main non-albicans species resistant to azoles class. However, in these times when emerging resistance is a worldwide concern we should expand our knowledge among other candida species resistant to theazole class. Therefore, we need to be aware of the resistance profile of Candida duobushaemulonii, an emerging pathogen in the healthcare system. In addition, candidemia as the cause of sudden deterioration in a critically ill patient and in patients requiring the use of central line catheters need to be always considered.

References


AMYOPATHIC DERMATOMYOSITIS AND RHEUMATOID ARTHRITIS: AN UNCOMMON PRESENTATION OF OVERLAP SYNDROME

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Introduction: Overlap syndrome is a term used to describe two or more systemic rheumatic diseases identified in the same patient, in which each disease fulfils its own diagnostic criteria. Inflammatory myopathies such as Dermatomyositis are associated with an increased risk of malignancy and interstitial lung disease. It may overlap with Systemic Sclerosis, Systemic Lupus Erythematosus, Mixed Connective Tissue Disease and less often Rheumatoid Arthritis. We presented a case of an atypical presentation of overlap syndrome with unremarkable labs findings.

Case Presentation: This is a case of 30 yo female who presented with violaceous eruption and edema on the upper eyelids (heliotrope rash). She visited emergency room few times and was diagnosed as an allergic skin reaction and treated with antihistamine medications without improvement of symptoms. Few months later she developed significant functional limitations marked by mechanic’s hands, Gottron’s papules, Gottron’s sign, perungual erythema with inflammatory arthritis and synovitis of the metacarpophalangeal joints, proximal interphalangeal joints, elbows and knees. Associated with fever episodes and alopecia without clinical evidence of muscle weakness. Laboratory data was significant for positive rheumatoid factor, anti-CCP antibodies, and normal CPK levels. Skin biopsy findings were typical of dermatomyositis with superficial and deep perivascular lymphocytic infiltrate.

Discussion: Overlap syndrome is when a patient meets diagnostic criteria for two or more connective tissue disease (CTD). About 25% of patients with CTD presents with overlapping clinical manifestations, more commonly in females than in males with a 9:1 ratio. About 11-40% of patients with DM present as an overlap syndrome.

Dermatomyositis (DM) is an inflammatory disease that includes characteristic skin rash, symmetric proximal muscle weakness and elevated skeletal muscle enzymes as described in Bohan and Peter’s criteria. Amyopathic DM is a subtype of DM characterized by biopsy-confirmed skin rash of classic DM for ≥6 months with no evidence of muscle weakness or elevated muscle enzymes as proposed by Sontheimer. This patient having typical skin rash of DM heliotrope rash, Gottron’s sign/papules; no proximal muscle weakness, normal CPK levels and fever meet the diagnostic criteria of amyopathic DM.

RA is the most common inflammatory arthritis involving ≥3 joints, positive RF and/or anti-CCP, disease duration of >6 weeks, and elevated CRP or ESR. This patient also meets the 2010 ACR/EULAR criteria for RA which represent an unusual case of overlap syndrome of ADM and RA.

Importance of recognizing and diagnosing amyopathic DM for an early diagnosis and treatment of associated complications like ILD. Recommended PFTs, chest X-ray and CT scan if symptoms like dyspnea and persistent non-productive cough are associated. Patients with MDA-5 antibody have higher risk of ILD, including rapidly progressive presentation and high mortality. Screening for internal organ malignancy. Early Dx and Tx of RA to prevent irreversible joint damage.

References
Introduction: Lung neuroendocrine tumors (NETs) are a rare group of pulmonary neoplasms with heterogeneous pathological features that account for approximately 25% of primary lung neoplasms. Within the four subtypes of pulmonary NETs, well-differentiated, low-grade typical carcinoids (TCs) account for only 2% percent of primary lung neoplasms. Clinically, they may be asymptomatic, present with non-resolving recurrent pneumonitis, hemoptysis or with paraneoplastic syndromes. We present a case of post obstructive pneumonia in a young patient who showed features of central bronchial typical carcinoid in high resolution computed tomography (HRCT) and later confirmed computerized tomography (CT) guided biopsy.

Case Presentation: A 36-year-old woman with past medical history of asthma presents to the office for evaluation of a lung mass discovered after an episode of pneumonia. Patient reports an episode of pneumonia that was initially managed with outpatient antibiotic therapy with no improvement. Patient later was hospitalized to complete intravenous (IV) antibiotics and was found on CT scan with a centrally located mass. CT guided biopsy was performed by interventional radiology with findings of well-differentiated carcinoid tumor with CD 56, Ki-67 and Synaptophysin C positivity. Six months later patient presents to our clinic for evaluation. Patient referred pleuritic chest pain with deep inspiration and no other associated symptoms. Physical examination showed right basal dullness with absent breath sounds. Imaging studies revealed right lower lobe mass with calcifications in infrahilar region measuring 3.7 x 4.8 cm. Bronchoscopy was necessary to identify exact location of the mass inside bronchial tree which would determine further surgical management. A round, vascular mass occluding the secondary bronchus with no visualization of right middle and lower lobe entrance was noted. Lesion located approximately 2 cm off secondary carina, between right upper lobe and middle lobe entrance. Bronchoscope was passed around lesion, demonstrating a mass that is evaginated inside bronchial tree arising from right lower lobe.

Discussion: A diagnosis of typical carcinoid is often overlooked in young patients with absence of specific symptoms that accompany lung NETs; clinical diagnosis is controversial and difficult. Apart from symptoms like chest pain, pleural effusion, cough, wheeze, hoarse voice, or atelectasis, patients may present with a pneumonia process. Recognition of nonspecific symptoms and variable natural disease history is key to accurate and timely diagnosis. Similarly, raising awareness of the increasing incidence of these tumors will lead to consensus in management guidelines and better prognosis.

References

QATAR - CLINICAL VIGNETTE POSTER FINALIST - MUHAMMAD JAMSHAID, MBBS

Bile Cast Nephropathy: A Case Report

Authors: Muhammad Bilal Jamshaid, Zohaib yousaf, M. Mohamedali, Adeel Ahmad Khan

Introduction: Bile cast nephropathy is characterized by deterioration of renal function in the presence of hyperbilirubinemia. Management guidelines do not exist due to few reported cases. (1,5)

While the exact cause is unknown, bile cast nephropathy is presumed to be secondary to multiple concurrent insults to the kidney including direct toxicity from bile acids, obstruction by bile casts and systemic hypoperfusion from vasodilation. Therapies aimed at decreasing bilirubin, dialysis and plasmapheresis have been used in the management with varied success. (2)

We are reporting a case treated presumptively as bile cast nephropathy, which responded to hemodialysis and PTC Drain.

Case Presentation: 64 years old gentleman with multiple comorbidities including CKD stage 2, presented with yellow sclerae and dark urine. He was found to have obstructive jaundice as investigation revealed dilated intrahepatic biliary radicles, common bile duct and a pancreatic head lesion. Blood work up revealed unremarkable cell count with predominantly direct hyperbilirubinemia & acute kidney injury*, which resolved with hydration. Biopsy of the head of pancreas revealed adenocarcinoma not amenable to surgery due to the high perioperative risks. The decision to proceed with ERCP and stenting was made while seeking second opinion from a high-volume center.

Patient developed hypotension and features suggestive acute cholangitis with raised inflammatory markers (WBCs 17, CRP 130, procalcitonin 3.2) before ERCP along with anuria and acute renal failure*. He was fluid responsive but remained anuric. He was initiated on antibiotics and PTC drain was inserted. Hemodialysis was initiated with a plan to proceed to plasma exchange in case of inadequate response. After 3 sessions of dialysis patient started to produce urine & renal function improved*. The concomitant worsening of renal function with hyperbilirubinemia & concomitant improvement of both following dialysis is highly suggestive of bile cast nephropathy. Potential contributing factors include ischemic ATN, and sepsis. Although renal biopsy is diagnostic but poses a challenge in this high-risk patient.

*refer to table

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<th>Post Dialysis/After PTC</th>
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Discussion: Hepatorenal syndrome is a common working diagnosis in patient’s acute kidney injury with a background of liver disease. Bile cast nephropathy is one of the main causes of AKI other than hepatorenal syndrome in these patients (1,3)

A clinicopathologic study from University of Chicago involving 44 subjects with jaundice & renal injury showed 24 patients had bile casts upon biopsy/autopsy. Eleven of the 13 patients labelled as hepatorenal syndrome had tubular bile casts. (3,4)

Bile cast nephropathy is an under-reported entity. It should be considered when encountering a patient with kidney injury in a patient with liver disease.

References

Hyponatremia caused by Acute Secondary Adrenal Insufficiency due to Pituitary Apoplexy

Authors: Russell Bratman MD, Fabio Lima MD, Indu Voruganti MS4, Ricardo Correa MD, Justin Yoon MD, Kristina W. Berglund MD

Introduction: Hyponatremia is a commonly-encountered clinical scenario which carries a broad differential diagnosis. We present a case of acute-on-chronic hyponatremia due to an unusual etiology, pituitary apoplexy resulting in acute secondary adrenal insufficiency.

Case Presentation: An 85-year-old female with a reported past medical history of chronic syndrome of inappropriate antidiuretic hormone (SIADH) on salt tabs, coronary artery disease, hypertension, and hyperlipidemia presented to the hospital with a one week history of otalgia and odynophagia and a one day history of emesis. Laboratory workup revealed hyponatremia to 117 mEq/L on a baseline of approximately 130 mEq/L. Serum osmolality was 248 mOsm/kg and urine osmolality was 590 mOsm/kg. On the medical service, SIADH treatment with fluid restriction and diuretic therapy were unsuccessful in correction of the serum sodium. Further workup was significant for a low AM cortisol (obtained 7AM) of 3.9 ug/dL, a normal thyroid stimulating hormone level (1.622 uIU/ml), and a normal cosyntropin stimulation test (3.0, 15.6, 20.6 ug/dl). Endocrinology was consulted and further history revealed a pituitary tumor that had been lost to follow-up; MRI of the pituitary was suspicious for pituitary apoplexy. Sodium improved to baseline rapidly after the administration of high-dose intravenous hydrocortisone. Free thyroxine level was found to be decreased to 0.65 ng/dl and the patient was given supplemental levothyroxine. The patient was transferred to the neurosurgical service and was managed nonoperatively.

Discussion: When adrenal insufficiency and hypothyroidism are considered in the differential diagnosis of hyponatremia, attention must be paid to the possibility of primary, secondary, and tertiary causes. SIADH and adrenal insufficiency can both result in hypotonic serum with hypertonic urine, so adrenal insufficiency should be considered if hyponatremia felt due to SIADH does not respond to therapy for same. The proposed mechanism is that corticotropin releasing hormone (CRH), the secretion of which increases in adrenal insufficiency, also leads to antidiuretic hormone (ADH) secretion. Adrenal insufficiency can result from primary (adrenal), secondary (pituitary), and tertiary (hypothalamic) causes. All such etiologies for adrenal insufficiency must be considered when the diagnosis is entertained. High-dose (250 µg) cosyntropin stimulation testing, while sensitive for primary adrenal insufficiency, is not sufficiently sensitive to rule out acute secondary causes. If adrenal insufficiency is suspected due to low AM cortisol and cosyntropin stimulation testing is normal, workup for secondary causes (such as with MRI brain) should be considered. Thyroid stimulating hormone level may be inappropriately normal in the setting of secondary hypothyroidism, so free T4 is the only reliable test.
RHODE ISLAND CLINICAL VIGNETTE POSTER FINALIST - DINA IBRAHIM, MD

A CASE REPORT OF RARE PROGRESSIVE STENO-OCCCLUSIVE SYNDROME: MOYAMOYA DISEASE

Authors: Dina A. Ibrahim MD, Ali Alkhayat MD, Ulviyya Gasimova MD, Ph.D, Salaheldin Elhamamsy MD, FACP, Affiliation: Kent Hospital / Brown University

Introduction: MoyaMoya disease is a rare disease, in which the patient in different age groups may suffer from refractory headaches and recurrent strokes, as a result of pathological deposition of fibrin on the walls of the intracranial arteries.

Case Presentation: A 50-year-old Caucasian male with medical history of diabetes mellitus type II, hypertension, hyperlipidemia, severe right Common Carotid Stenosis and transient ischemic attacks, presented to our primary care office. His previous medical history also included stroke in territory of right Middle Cerebral Artery (MCA) and thoracic spinal cord infarction in 2009 with subsequent residual left-sided weakness.

In June 2016, a CT angiogram of the head showed stenosis within the right intracranial internal carotid artery (ICA) with chronic occlusion of the M1 segment. A collateralized flow was visualized as well. A CT angiogram of the neck revealed a plaque and calcification at the right carotid bulb with resultant marked narrowing (>90% occlusion) in the proximal right ICA with resultant pinpoint flow in the occluded area. Moreover, an atherosclerotic plaque and calcification were seen in the left carotid bulb with approximately 50-69% stenosis in the proximal left ICA. MRI of the brain also showed multiple chronic infarcts within the right MCA-ACA and MCA-PCA watershed distributions of Circle of Willis. These findings were the same when compared to the results of radiological studies done back in 2011. These finding opted later on for the diagnosis of MoyaMoya disease.

Discussion: MoyaMoya disease is a chronic, progressive occlusion of the Circle of Willis arteries that leads to the development of characteristic collateral vessels seen cerebral angiography. MoyaMoya was originally considered to affect predominantly persons of Asian heritage but has now been observed throughout the world. The incidence peaks lie within two age groups: children who are 5 years old and adults in their mid-40s. It is however rarely seen in the African American population. Pathologically, MoyaMoya is associated with fibrin deposition in the walls of watershed area arteries of Circle of Willis, which leads to proliferation, and occlusion causing the clinical scenario of recurrent strokes. It usually presents with recurrent headaches and is migraine like in quality and refractory to medical therapies. Although MRI angiography is used to confirm the diagnosis, CT angiography can also be used to see intracranial stenoses suggesting MoyaMoya. Thus, CT angiography can be considered when MRI is not readily available and a diagnosis of cerebral occlusive vasculopathy is being considered. Surgery is the only viable option till date. A good option for adult symptomatic patients is superficial temporal artery-middle cerebral artery bypass or middle meningeal artery to middle cerebral artery bypass. Patients were reported to have 96% probability of remaining stroke free over the subsequent five years.

References


Introduction: Antiphospholipid Antibody Syndrome (APS) is an autoimmune induced thrombophilia defined as the occurrence of vascular thrombosis or pregnancy loss with the presence of antiphospholipid antibodies. One percent of cases of APS are characterized as Catastrophic APS (CAPS) with multiorgan failure due to thrombosis that can often mimic sepsis. This case illustrates the need to understand the diagnosis and treatment CAPS with particular attention to measuring anticoagulation in the presence of lupus anticoagulants and occult malignancy.

Learning Objectives

1. Recognize signs and symptoms of Catastrophic APS.
2. Learn to quantify adequate anticoagulation in APS by using a chromogenic factor X.
3. Recognize the perils of premature closure bias in patients with undifferentiated systemic disease.

Case Presentation: A 46-year-old man with a history of APS complicated by pulmonary embolism (PE), chronic thromboembolic lung disease presented with fever, chills, malaise, dyspnea and diffuse progressive subcutaneous nodules after handling fish. On admission, patient was newly hypoxic to 90% and required supplemental oxygen at 5 liters. Chest CT angiography showed multifocal areas of consolidation, left upper lobar PE, and pleural effusions. His INR was 2.2 at admission, so coumadin was continued at his home dose. He was also noted to have cellulitis in his thigh and multiple scattered subcutaneous nodules, most prominently in the right lower quadrant. He was initially started on vancomycin, piperacillin-tazobactam, and azithromycin. Despite antibiotics and aggressive diuresis, his hypoxia progressed and required transfer to the ICU for mechanical ventilation. He developed multiorgan failure and eventually passed away. An autopsy was completed with gross lung pathology revealing left upper and lower lobe PE and multifocal necrotic nodules comprised of anaplastic large cell lymphoma. Pathology did not show pneumonia or mycobacterium infection.

Discussion: This case is important to all physicians who provide care in the hospital because it demonstrates the need to use accurate testing to effectively manage catastrophic antiphospholipid antibody syndrome; a dangerous disease with high mortality. In this case, the patient’s immunosuppression, marine exposure and nodules initially suggested the presence of a mycobacterial infection, prompting an infectious disease workup. Patients with APS can have a falsely elevated INR, which makes monitoring of anticoagulants very challenging, particularly in the setting of active thrombosis. Chromogenic Factor X is preferred for anticoagulant monitoring. In this case, it is likely that the development of PE was influenced by both APS and the occult malignancy; which mimicked pneumonia on lung imaging. This case underscores the importance of accurately measuring coagulation in APS and the potential need for advancing anticoagulation despite normal INR. The case also demonstrates the need to avoid premature closure when evaluating worsening hypoxia in an immunosuppressed patient with APS. Nonetheless, the presence of severe multiorgan invasion of anaplastic large cell lymphoma portends a grim prognosis from the outset.

References


SOUTH DAKOTA CLINICAL VIGNETTE POSTER FINALIST - RADOWN ELNAIR, MD

Statin-associated Necrotizing Autoimmune Myopathy (NAM): Sore muscles and a painful memory

Authors: Radowan Elnair MD, Joseph Fanciullo MD, Department of Internal Medicine, University of South Dakota Sanford School of Medicine

Introduction: An estimated 39.2 million Americans are on a statin. Statins may cause muscle-related symptoms in 10 to 20% of patients, but these symptoms usually resolve within weeks after the medication is stopped. Autoimmune myositis is rare, with an estimated prevalence of 22 in 100,000 and statin-induced Necrotizing Autoimmune Myopathy (NAM) is rarer still. Symptoms include muscle weakness and myalgia, although more profound muscle weakness with respiratory failure can also occur. Marked elevation in CK is characteristic.

Case Presentation: A 77-year-old female with past medical history of diabetes, hyperlipidemia, and vitamin D deficiency developed progressive diffuse weakness over the course of a year. She had no myalgias, skin rash, dysphagia, Raynaud's phenomenon, shortness of breath, or neuropathic pain. She had a remote history of smoking over 30 years ago and carried no diagnosis of cancer. Physical examination was unremarkable, apart from symmetric weakness in the proximal muscle groups in both upper and lower extremities (power 4/5), with no evidence of muscle tenderness, synovitis or skin changes. Initial evaluation by her primary care physician showed an elevated CK level. This led to discontinuation of atorvastatin, however CK levels failed to normalize, which warranted further workup. Testing for 3-hydroxy-3-methylglutaryl-coenzyme A Reductase (HMGCR) antibodies was positive. CT of the abdomen and pelvis revealed no evidence of malignancy. She agreed to be evaluated by rheumatology approximately 1 year after discontinuation of atorvastatin. At the time, she showed no limitation in her activities of daily living. Given the constellation of symptoms and laboratory findings; skeletal muscle biopsy was recommended. A tissue sample from the left proximal thigh showed ongoing mild necrotizing myopathic changes. Without evidence of myositis; the findings along with history of positive HMGCR antibody testing were consistent with a diagnosis of statin-induced NAM. Given her age and the potential for serious adverse reactions with immunosuppression; she elected for management with an expectant approach. She continues to have an elevated CK level but remains independent in her activities of daily living twenty months after the diagnosis was established.

Discussion: Overall, up to 20% of patients will develop statin related myalgias. The pathophysiology of NAM is poorly understood. Nearly all patients with statin-induced NAM will have positive anti-HMGCR antibodies, which are absent in most patients with self-limited statin myopathy or in healthy controls. Along with avoidance of all statins, immunosuppression is often needed and is usually given in the form of corticosteroids initially, followed by longer-term systemic immunosuppression. The condition tends to respond favorably to treatment, with improvement in muscle strength and reduction of CK levels. In most cases; anti-HMGCR levels do not return to the normal range even in those who have clinical remission. Longer-term management of hypercholesterolemia remains a challenge in patients who have suffered from statin-induced NAM.

References

LGL Leukemia and Auto-immunity: Clinical Clues to Avoid Diagnostic Delay

Authors: Alexandra M. Haugh, MD, MPH, Shannon Stockton, MD, Peter Edmonds, MD, Jiun-Ruey Hu MD, MPH, Claudio A. Mosse MD, PhD, Brian W. Christman, MD

Introduction

Case Presentation: A 69-year-old man presented to the ED with pain and swelling on the dorsum of his right hand. He had been seen twice in the preceding two weeks for the same symptoms but returned with worsening edema, erythema and fever to 101.9F despite 10 days of antibiotic therapy. Physical exam findings included extensive erythema and edema of his right hand as well as subluxation at the MCP joints bilaterally. Laboratory evaluation revealed a WBC of 1.16 and an ANC of 0, prompting treatment with broad spectrum antimicrobials for cellulitis with neutropenia. Greater than 80% of the WBC on his peripheral smear were large granular lymphocytes with few, if any, neutrophils noted.

The patient had a history of rheumatoid arthritis, Hepatitis C and Large Granular Lymphocytic Leukemia (LGLL), which was diagnosed in 2014 following several cutaneous abscesses and diverticulitis with an enterocutaneous fistula. Treatment with cyclophosphamide resulted in a complete remission in 2015. After two days of IV antibiotics and rapid improvement in his symptoms, he was discharged with plans for outpatient chemotherapy.

Discussion: LGLL is relatively rare, representing 2-5% of chronic lymphoproliferative disorders in North America. An estimated one-third of patients are asymptomatic at the time of diagnosis but most patients present with aphthous ulcers or bacterial infections involving the skin, oropharynx or perirectal areas. LGLL is also associated with several auto-immune disorders, most commonly with rheumatoid arthritis, which is reported in 10-18% of patients.

LGLL is thought to be driven by unknown antigen stimulation, which first drives the formation an oligoclonal cytotoxic lymphocyte population. Chronic stimulation promotes dysregulation of apoptosis and ultimately leads to the expansion of a dominant lymphocytic clone. Growth of this monoclonal cytotoxic T-cell population triggers the production of inflammatory cytokines and the release of cytotoxic granules by NK cells, resulting in fatigue, anemia, neutropenia and inflammation.

Given the constitutive T-cell activation seen in LGLL, immunosuppressive therapy is first line treatment. Indications for treatment include severe neutropenia, moderate neutropenia with recurrent infections and symptomatic anemia. While the prognosis of LGLL is generally better than that seen with many other adult leukemias, complete response rates to therapy are low and relapse should be expected. Most patients, however, will continue to respond to immunosuppressive therapy, which often can be continued indefinitely.

Our patient suffered from numerous episodes of skin and soft tissue infections and febrile neutropenia before the original diagnosis of LGLL. Despite a robust response to cyclophosphamide at diagnosis, he ultimately presented with complicated cellulitis secondary to disease recurrence and will require further immunosuppressive therapy. Although rare, prompt recognition and treatment of this disorder can often prevent many of the associated infectious and pancytopenic complications.

References


pg. 631
TWICE BITTEN: A RARE CASE OF HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS ASSOCIATED WITH EBV AND EHRlichia CHAFFEENSIS

Authors: Mary Gadalla M.D., Chris Pasvantis M.D., Steve Embry M.D.

Introduction: Hemophagocytic lymphohistiocytosis (HLH) is a rare, aggressive, and life-threatening syndrome of excessive immune activation in which cytotoxic cells and macrophages hyper-proliferate causing rapid and severe systemic inflammation, multi-organ damage, and hemophagocytosis. Many patients with HLH have a predisposing genetic defect and/or an immunologic trigger that can include infection, malignancy, rheumatologic disease, and immunosuppressive drugs. It is most commonly seen in infants and young children but can rarely affect patients well into adulthood.

Case Presentation: The patient is a 57-year-old female with history of breast cancer in remission and rheumatoid arthritis on infliximab and sulfasalazine who presented to the ED for evaluation of several weeks of progressive malaise, dyspnea, and altered mental status. Initially, she was mildly hypoxic with bilateral infiltrates on chest x-ray and started on empiric antibiotics, including doxycycline, for pneumonia. Labs were notable for mildly elevated creatinine, ferritin, liver function tests, and mild pancytopenia. Initially stable, she exhibited rapid deterioration over the next 24 hours with hypoxic respiratory failure, acute liver injury, acute kidney injury, pancytopenia with neutropenia, and worsening encephalopathy and fever. She was eventually intubated and placed on pressors. Preliminary bone marrow biopsy results revealed significant hemophagocytosis prompting evaluation for HLH. She met diagnostic criteria with fevers, cytopenia, elevated triglycerides, elevated ferritin, and evidence of hemophagocytosis. High-dose steroids were started. Lumbar puncture revealed elevated protein and 345 WBCs, with lymphocytic predominance. Meningitis panel and CSF cultures were negative. Serum PCR for Ehrlichia chaffeensis and EBV IgM were positive; history of tick exposure was unknown. The next day, she developed worsening AKI. Troponin was elevated and TTE showed multiple wall-motion abnormalities and reduced EF. Her condition worsened despite steroids and antibiotics and her family chose to withdraw care. She expired four days after admission. Ehrlichiosis and EBV in the setting of infliximab use were thought to be the underlying trigger for her HLH.

Discussion: Secondary hemophagocytic lymphohistiocytosis (HLH) is a rare, but often fatal disease. Clinicians should have a high index of suspicion of HLH when encountering a case of septic shock with multi-organ failure and CBC abnormalities, especially if a source of sepsis is not identified. Ferritin level, though nonspecific, can be used in the initial diagnostic workup. A high ferritin level can lead to further laboratory evaluation, earlier diagnosis and prompt treatment, thereby leading to an improved survival. Also, the use of infliximab in this case is important as infliximab has been reported in certain cases of immunosuppression (SLE, RA) to treat refractory HLH. However, though anti-TNF-α therapy does not appear to be a direct trigger of HLH, it is a permissive source of severe infection that can lead to HLH due to immunosuppression.

References

TENNESSEE CLINICAL VIGNETTE POSTER FINALIST - DEEPANJALI R NAIR, MD

A Case of Monomac Syndrome Diagnosed in a 48-Year-old Female who Presented with Fever of Unknown Origin and Recurrent Episodes of Pneumonia

Authors: Deepa Nair (Department of Internal Medicine), Aaron Eskind (Department of Internal Medicine)

Introduction: Heterozygous mutation in the GATA-2 gene produces a loss of gene function, leading to haploinsufficiency1. This condition is called Monocytopenia and Mycobacterial infection (MonoMAC) syndrome. Patients will be immunodeficient, with marked susceptibility to human papilloma virus (HPV) and nontuberculous mycobacteria (NTM), predisposition to MDS/AML, pulmonary alveolar proteinosis and congenital lymphedema1.

Case Presentation: A 48-year-old Caucasian female presented to Saint Thomas West Hospital with fever of unknown origin. At the time of presentation, she was having intermittent fevers for the previous 5 weeks, associated with night sweats, chills, myalgias, shortness of breath and erythematous nodules on the anterior aspect of the shins. During the course of 5 weeks, she was treated with antibiotics and high-dose steroids. Her symptoms improved, but reappeared following taper of the steroids. Her past medical history was significant for recurrent episodes of pneumonia, bronchiectasis, Mycobacterium Avium Intracellulare infection (MAI) of bone marrow, squamous cell carcinoma of the vagina and vulvar dysplasia. On arrival, she was febrile up to 103.1 and tachycardic. Her chest had a non-erythematous nodular lesion and auscultation revealed bilateral fine crackles. Labs were notable for elevated white count with 91% neutrophils, 2% lymphocytes and 0% monocytes/eosinophils and elevated ESR and CRP. Immunoglobulin panel showed IgG subclass 2 deficiency. High-resolution CT of the chest showed emphysema and bronchiectasis in the mid-upper lung zones and small nodules in the right lower lobe. Extensive workup for infection, malignancy and vasculitis were negative. Punch biopsy of the nodule on the chest was positive for HSV-2 on viral culture. Due to her constellation of symptoms including MAI infections, genital dysplasia, genital warts, HSV-2 infection of the skin, recurrent pulmonary infections, IgG 2 deficiency and persistent monocytopenia, concern was raised for MonoMac syndrome. Genetic testing confirmed the diagnosis of GATA-2 deficiency.

Discussion: While initial clinical presentations vary, early diagnosis can dramatically alter the disease course. In one study, 48% of patients noted to have this deficiency died of causes ranging from malignancy to myelodysplasia, with the mean age of death at 34.7 years.2 However early recognition and referral for allogenic transplant can be lifesaving, as it may reverse the disease progression. It is therefore paramount that physicians include MonoMAC in the differential diagnosis when a patient presents with recurrent nontuberculous mycobacterial, viral and invasive fungal infections. Case reports such as this offer improved insight into the multiple manifestations of this rare disease entity.
NON-ALCOHOLIC WERNICKE ENCEPHALOPATHY IN THE SETTING OF HYPEREMESIS GRAVIDARUM AND POOR DIETARY INTAKE

Authors: Raissi, Azadeh (Internal Medicine/UT/Nashville); Doering, Tracey (Internal Medicine/UT/Nashville)

Introduction: Wernicke encephalopathy (WE) is an acute syndrome requiring emergent treatment to prevent death and neurologic morbidity. Although WE is most often associated with chronic alcoholism, it also occurs in other settings such as poor nutrition caused by malabsorption, poor dietary intake, and ESRD pts on dialysis. Autopsy studies have consistently revealed a higher incidence of Wernicke lesions in the general population than is predicted by clinical studies, suggesting that it is clinically under-recognized. WE produces petechial hemorrhagic necrosis in midline brain structures and corresponding deficits in mentation, oculomotor function, and gait ataxia. Only 1/3 of these pts have all 3 symptoms. Absence of one or more of the classic symptoms may lead to under-diagnosis. Thiamine replacement is mandatory early in disease.

Case Presentation: The patient is a 32-year-old non-alcoholic female who had an elective abortion due to severe hyperemesis gravidarum 3 weeks prior to presentation. Prior to the abortion, pt had anorexia, and weight loss. One week prior to presentation, she became minimally verbal with decreased eye contact, and developed short term memory loss. Psychiatry was consulted and diagnosis of major depressive disorder with catatonic features was made and olanzapine initiated for mood stabilization. Two days later, neurology consulted due to persistent symptoms. Nystagmus and intermittent myoclonic type jerks were noted on examination, and EEG was obtained which was normal. Neurologist agreed that the presentation was likely psychiatric in origin. Myoclonic jerks and nystagmus worsened and MRI was obtained five days after admission which revealed hyperintense T2/FLAIR signal abnormality involving the medial border of the thalami along the third ventricle extending to the mammillary bodies and to the periaqueductal gray regions consistent with Wernicke Encephalopathy. She was placed on high-dose thiamine and her signs and symptoms including oral intake, eye contact and verbal communication improved. She remained weak, unsteady and continued to have short term memory impairment. Patient was discharged home on oral thiamine. On return one month later, weakness, nystagmus, and ataxia had resolved. Three months after hospital discharge, EEG was repeated, with normal result. MRI was repeated and showed nearly complete resolution of lesions. Short term memory remained limited.

Discussion: The main barrier to diagnosis of WE is a low index of suspicion in the non-alcoholic patient, especially when the classic triad of clinical symptoms is not present and the patient is not a known alcoholic. Physician should have high index of suspicion for Wernicke encephalopathy for all at-risk patients with undiagnosed altered mental status, oculomotor disorders, or ataxia. Diagnostic testing should not delay treatment, which should immediately follow consideration of the diagnosis. Intravenous administration of thiamine is safe, simple, inexpensive, and effective in treating this condition.

References

Chromobacterium violaceum sepsis as a consequence of hurricane heroism and severe neutrophil deficiency

Authors: J. Patrik Hornak, MD; Alyssa L. Anderson, MD; Daniel A. Ortiz, PhD; Lola Carrete, MD; Lindsay K. Sonstein, MD; A. Clinton White, MD

Introduction: Chromobacterium violaceum is a catalase/oxidase-producing Gram-negative rod ubiquitous in tropical/subtropical regions that rarely causes clinical infections. Less than 200 cases have been reported globally, often in immunocompromised individuals. Mortality rate exceeds 60%. Usually, fulminant sepsis is the initial clinical presentation and the organism is commonly multi-drug resistant. Here we describe a dramatic case of C. violaceum bacteremia following floodwater exposure in a patient with neutrophil deficiency caused by an exceedingly rare, atypical variant of glucose-6-phosphate dehydrogenase deficiency (G6PDD).

Case Presentation: Three decades ago, a 3 year old boy with G6PDD—Beaumont subtype suffered a rapidly fatal infection with C. violaceum. His surviving twin also had G6PDD—Beaumont and was found to have < 5% neutrophil activity. Following Hurricane Harvey floodwater exposure, the surviving twin developed a cutaneous abscess and bacteremia due to C. violaceum. Admitted with sepsis and hemolysis, he was initially treated with vancomycin. Upon receipt of culture data, therapy was changed to aztreonam, gentamicin, and metronidazole without improvement. Both pigmented and non-pigmented phenotypes were cultured. With ongoing hemolysis, he was transferred to our center for possible splenectomy. Computed tomography (CT) showed hepatosplenomegaly with splenic infarct and innumerable pulmonary and hepatic septic foci. Based on available susceptibility data he was switched to meropenem, alas without further improvement. Though he previously described moxifloxacin as causing hemolysis, literature review showed fluoroquinolones to be the most bioactive agents, in vitro. Cautiously, levofloxacin was added to his regimen. Without hemolysis, his fevers and anemia steadily improved over the next two days. Meropenem was stopped and levofloxacin continued on discharge. He completed five of six planned weeks of therapy which was stopped due to diffuse arthralgias attributed to quinolone use. Repeat CT demonstrated resolution of the pulmonary and hepatic nodules and splenic infarct improvement.

Discussion: Historically, C. violaceum infection has been marked by frequent mortality, recurrence, and treatment failure. The organism shows high levels of resistance to penicillins and cephalosporins with variable susceptibility to beta-lactam/beta-lactamase inhibitors in a pattern consistent with extended-spectrum beta-lactamase (ESBL) production. Isolates remain susceptible to carbapenems, fluoroquinolones, and tetracyclines. Interestingly, culturing both pigmented and non-pigmented isolates has not previously been described clinically; stressed isolates have been shown to lose pigment-producing abilities. Additionally, this is the first report of C. violaceum infection involving a set of identical twins, the first in an adult with G6PDD, and the second case ever reported in Texas. Lastly, our case supports emerging evidence that fluoroquinolone-associated tendon toxicity could relate to increased reactive oxygen species production in chondrocytes, as our patient did not have other risk factors for fluoroquinolone-induced tendinopathy. RECENT UPDATE! Nearly one year following primary infection, our patient was hospitalized for acute cholecystitis and noted to be growing Chromobacterium violaceum on blood culture. Thus, the story continues...

References

Bleeding Due To Acquired Dysfibrinogenemia As The Initial Presentation In Multiple Myeloma

Authors: Namrah Siddiq, M.D., Colin Bergstrom, M.D., Srikanth Nagalla, M.D.

Introduction: Patients with multiple myeloma (MM) are at risk for acquired dysfibrinogenemia due to interference by paraproteins in the coagulation pathway resulting in laboratory abnormalities and/or bleeding complications. Generally, patients are diagnosed with MM prior to the discovery of dysfibrinogenemia. We describe a unique case of bleeding diathesis which after work-up revealed the presence of dysfibrinogenemia, leading to an underlying diagnosis of MM.

Case Presentation: A 63-year-old man with a history of coronary artery disease presented to our hospital for work up of bleeding diathesis. He was previously admitted to an outside hospital with a hemoglobin of 6 g/dL and CT findings revealing a new peri-pancreatic hematoma. The patient also had a recent history of easy bruising, prolonged bleeding with shaving, and hematuria. Physical exam demonstrated the presence of ecchymoses on his extremities. Lab work on admission was notable for leukocytosis and mild anemia. He was also found to have an elevated protime (PT), borderline elevated prothrombin time (PTT), a low fibrinogen level, a normal fibrinogen antigen level, prolonged thrombin time, slightly prolonged reptilase time, normal vWF, and minimally decreased Factor VII, V, and X activity. The results of serum protein electrophoresis (SPEP) showed 0.11 g/dL oligoclonal bands with a kappa to lambda ratio of 0.05. Bone marrow biopsy revealed 20-25% lambda light chain restricted plasma cells. MRI was notable for two sacral ala lytic lesions. These collective findings suggested a diagnosis of multiple myeloma. The patient was refractory to initial treatment with Bortezomib/Dexamathasone, indicated by a decrease in fibrinogen, increase in lambda free light chains (LFLC), and recurrent bleeds. This prompted a switch to Carfilzomib/Dexamethasone, leading to an improvement in LFLCs and normalization of fibrinogen. The patient eventually underwent definitive therapy with an autologous peripheral blood stem cell transplant.

Discussion: Multiple myeloma has been associated with acquired dysfibrinogenemia. The mechanism is thought to be secondary to inhibition of fibrin polymerization by paraproteins. Our patient had a normal fibrinogen antigen level but an abnormally low fibrinogen activity level, which raised concern for dysfibrinogenemia. Other case reports have highlighted patients with a prior diagnosis of multiple myeloma who then presented with acquired dysfibrinogenemia based on lab abnormalities or bleeding tendencies. However, our case discusses a patient presenting with bleeding diathesis and dysfibrinogenemia, which after further work-up led to a diagnosis of multiple myeloma. Recurrent bleeds are a unique presentation for undiagnosed multiple myeloma. Therefore, in middle aged and elderly patients with recurrent bleeds and abnormal fibrinogen activity, multiple myeloma should be included in the differential. Further lab studies should include an SPEP and serum free light chains. Previous cases have reported that in patients with MM and dysfibrinogenemia, treatment with chemotherapy or plasma exchange resulted in resolution of symptoms. Our case had a novel clinical course characterized by a refractoriness to initial therapy requiring a change in treatment.

References


Calcified Amorphous Tumor presenting as Syncopal Episode

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Introduction: Calcified amorphous tumor of the heart is a rare non-neoplastic cardiac mass that may mimic malignancy on imaging. The mass is characterized by nodular calcium in the background of amorphous degenerating fibrinous material. Clinical diagnosis is extremely difficult and current imaging techniques do not readily differentiate cardiac CAT from other cardiac lesions including myxomas, cardiac thrombi, or other vegetations. Surgical resection is the standard approach for both diagnosis and treatment for prevention of obstructive symptoms and embolization.

Case Presentation: A 68 year old female with no cardiac history presented to our emergency department after a syncopal episode along with dyspnea on exertion and bilateral lower-extremity swelling. X-ray showed pulmonary vascular congestion with trace pleural effusions. Transthoracic echocardiography showed normal systolic and diastolic function but revealed a mobile, echogenic mass attached to the ventricular surface of the anterior mitral valve leaflet. A subsequent transesophageal echocardiogram confirmed a highly mobile 20 mm x 7 mm mass attached to the anterior mitral valve annulus, prolapsing into the left ventricular outflow tract. The clinical differential diagnosis consisted of cardiac neoplasm (myxoma, fibroelastoma), sterile vegetation, or thrombus. The patient had no signs or symptoms of endocarditis and the mass presented on the ventricular side of the mitral valve, so we felt it was less likely to be a septic vegetation. The patient eventually underwent resection of the intracardiac mass. Microscopic examination showed nodular calcified amorphous debris with admixed degenerated fibrin and focal chronic inflammation confirming the diagnosis of calcified amorphous tumor.

Discussion: First described 1997, cardiac CAT is extremely rare with very few cases being reported in available literature. The patients are typically asymptomatic, but when present, features of obstruction including dyspnea and syncope predominate. The pathological characteristics of CAT are nodular deposits of calcium in a background of eosinophilic amorphous materials and fresh fibrin. These clinical and pathological findings were identical to our case. Cardiac CAT has an excellent prognosis following surgical removal. Diagnosis based on imaging alone is difficult, hence surgical resection and histopathological examination is the sole modality for diagnosis. Cardiologists, CT surgeons, and pathologists should be aware of this entity in the differential diagnosis of a cardiac mass.
Is it Valve Vegetation, Thrombus, Myxoma or Something Mimicking All of These?

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Introduction: The differential diagnosis of intracardiac masses is a broad, diagnosing the mass type is challenging but crucial to determine the management and prevent complications. Three principle intracardiac masses are present; tumor, thrombus, and vegetation. Our patient presented with an embolic stroke and intraventricular mass seen on imaging attached to the mitral valve.

Case Presentation: A 70-year-old man with history of Diabetes Mellitus and hypertension presented with vertigo for one week. The patient was a febrile, normotensive and having normal regular heart rate. His physical exam was significant for holosystolic murmur radiated to the axilla, left-sided drift, and dysmetria. MRI of the brain revealed a small multifocal stroke concerning for embolic source. Transthoracic and transesophageal echocardiography showed severe mitral annular calcification (MAC) and a 1cm by 1cm "popcorn morphology" mobile mass attached by a stalk to the ventricular side of the posterior mitral leaflet. Repeated multiple blood cultures were negative in this patient. A coronary angiogram revealed 80% stenotic left anterior descending artery. The patient then underwent a concomitant coronary artery surgery and mass excision surgery. A histopathological examination revealed a very rare pathology of calcified nodular histiocytic proliferation, consistent with a reactive or reparative process secondary to MAC.

Discussion: MAC is a common diagnosis in the elderly population which is characterized by progressive calcium deposition along and below the mitral valve annulus. Our patient presented with a mass that was attached to the mitral valve which poses a diagnostic dilemma hence the rare presentation of MAC causing mass, and the resemblance of this mass to other more common diagnoses like valve vegetations, thrombosis or myxoma. The morphology of the mass (popcorn morphology) was favoring the diagnosis of atrial myxoma over valve vegetation; furthermore, repeated blood cultures in this patient were negative, and the patient did not meet Duke’s criteria for infective endocarditis. MAC was noticed by echocardiography which thought to be an incidental finding initially. The surgical decision was made based on the presumed diagnosis of myxoma based on morphology. MAC was confirmed with a histopathological examination after surgical excision of the mass. MAC has been shown to increase the risk of stroke, but it is unclear if the mass presentation will result in an added risk of stroke to patient presented with MAC. Physicians should keep a wide differential for patients who presented with intracardiac masses and MAC. MAC should be suspect as a possible cause of intracardiac mass attached to the mitral valve.
Overlap syndrome: Lupus and ANCA vasculitis presenting as pulmonary-renal syndrome

Authors: Rani Bhatia MD, Kartik Valluri MD, Kalpalatha Guntupalli MD

Introduction: Pulmonary-renal syndrome is a broad disease category with many distinct entities, of which systemic lupus erythematosus and antineutrophilic cytoplasmic antibody (ANCA)-associated vasculitis (SLE/AAV) overlap syndrome is a fairly new addition. Atypical symptoms occurring in the course of lupus, including intra-alveolar hemorrhage, should prompt evaluation for both ANAs and ANCAs.

Case Presentation: A 43 year-old-woman with no known medical history presented with two months intermittent fever, cough, pleurisy, hematuria, and weight loss. On exam she had diffuse crackles and lower extremity edema. The patient was intubated for respiratory distress. Laboratory workup revealed acute anemia and urinalysis consistent with nephritic syndrome. Chest x-ray showed bilateral diffuse opacities and subsequent CT scan showed centrilobular ground-glass opacities and consolidation. Bronchoscopy was indicative of diffuse alveolar hemorrhage. The patient also developed rapidly progressive renal failure and was started on pulse dose steroids. Autoimmune evaluation showed ANA 1:320 speckled pattern, MPO-ANCA+, and PR3-ANCA+. Renal biopsy revealed class IV diffuse lupus crescentic nephritis as well as necrotizing crescentic glomerulonephritis consistent with a concomitant ANCA-associated glomerulonephritis. The patient received five sessions of plasma exchange with nearly complete resolution of symptoms and was discharged on a slow prednisone taper.

Discussion: Pulmonary-renal syndrome (PRS) is a combination of diffuse alveolar hemorrhage and rapidly progressive glomerulonephritis. SLE/AAV overlap syndrome is a fairly new entity in the PRS category that usually has a severe initial presentation, with both vital and functional prognoses being poor. When managing a patient with diffuse alveolar hemorrhage and suspected lupus, awareness of this syndrome should prompt a thorough and early evaluation, as these patients have presence of both ANA and anti-MPO antibodies. Treatment includes corticosteroids and cytotoxic agents. Early initiation of plasma exchange has been shown to improve renal recovery and decrease progression to ESRD, although the long-term benefit of mortality remains unclear.
Typhus-induced Atrial Fibrillation with Rapid Ventricular Response

Authors: J. Michael Blair MD, Baylor College of Medicine

Introduction: Murine typhus is a febrile illness caused by *Rickettsia typhi*, transmitted by fleas (most commonly from rats). In temperate regions, the disease is most common during hot, dry periods. It can be misdiagnosed due to nonspecific symptoms. The classic triad is fever, rash, and headache; observed in 33-59% of cases.

Case Presentation: 68-year-old man presented during summer with two weeks of weakness and shortness of breath. He had associated chills, myalgia, arthralgia, blurry vision, dry mouth, dizziness, dry cough, and dysuria. On initial presentation, he had new-onset atrial fibrillation (AFib) with rapid ventricular response and hypotension prompting admission to intensive care. After treatment with amiodarone and metoprolol he converted to normal sinus rhythm and was started on apixaban. He was transferred to Medicine service and continued to have fatigue, myalgia, weakness, dry mouth, and subjective fevers. He had extensive animal exposure including two dogs, one cat, and a parrot at home. He lived in Texas by the bayou, known to have a lot of rats. He noted fleas on himself in the past. On exam, he appeared ill and fatigued with significant shortness of breath. Conjunctiva were slightly injected, there was marked xerostomia, and cervical lymphadenopathy. His pulse was normal rate and regular rhythm. There were normal heart sounds without murmurs. Slight bibasilar crackles heard on lung auscultation. There was vague abdominal tenderness. Skin appeared mottled. Labs were notable for thrombocytopenia, leukocytosis with neutrophilic predominance and left shift, elevated procalcitonin, hyperbilirubinemia, elevated liver enzymes, and elevated lactate dehydrogenase. Peripheral smear had atypical lymphocytes without bacterial or fungal inclusions. Tests for influenza, RSV, HIV, Bartonella, mononucleosis, Cryptococcus, Legionella, viral hepatitis, and Histoplasmosis were negative. Blood, urine, and sputum cultures were negative. Thyroid function tests were normal. Chest X-ray revealed mild interstitial edema and computed tomography showed airspace and nodular opacities in the lung bases. He was empirically started on doxycycline given concern for zoonotic bacterial infection. He continued to feel poorly and had worsening hepatocellular injury for two more days, after which he started to improve significantly. Eventually typhus immunoglobulin (Ig)M and IgG antibodies resulted strongly positive. On discharge, he felt markedly better, and all labs were improving. He has paroxysmal AFib that had not returned at follow-up and is presumably the result of murine typhus.

Discussion: The syndrome of a febrile illness with a prominence of headache and skin changes, thrombocytopenia, and liver injury was concerning for zoonotic bacterial infection including rickettsia or similar intracellular bacteria (e.g. rocky mountain spotted fever, ehrlichiosis, murine typhus, leptospirosis, etc). Empiric treatment with doxycycline is recommended for patients in whom a diagnosis of murine typhus is suspected. The illness is often self-limited however antibiotics significantly shortens duration.

References

When Blood Pressure Becomes too High: A Rare Case of Hypertensive Emergency Induced Thrombotic Microangiopathy in a Pregnant Patient

Introduction: Hypertensive emergency is defined as elevated blood pressure with organ damage. Hypertensive emergency in pregnant patients can cause many serious complications to both the mother and the fetus and are often commonly associated with eclampsia and HELLP syndrome. The presentation of HELLP syndrome can be difficult to distinguish from thrombotic microangiopathies (TMA) caused by conditions such as thrombotic thrombocytopenic purpura (TTP) and hemolytic uremic syndrome (HUS). Apart from TTP and HUS, another rare cause of TMA is hypertensive emergency. Here we present a rare case of TMA caused by hypertensive emergency in a pregnant patient.

Case Presentation: A 35 year-old pregnant African-American female at 27 weeks gestation presented to labor and delivery with dyspnea for 2 days. Patient had medical history of gestational hypertension and little prenatal care. Patient was noted to have a blood pressure of 263/140 and was admitted for further management. IV labetalol was administered without improvement and the decision was made to perform emergent C-section due to fetal distress. Post-operatively, patient was unable to be extubated and was transferred to ICU. Chest x-ray showed severe diffuse bilateral pulmonary edema. Patient’s labs were significant for hemoglobin 9.4 g/dL, hematocrit 28.0%, platelet 64,000, creatinine 7.71 mg/dL, blood urea nitrogen 80 mg/dL, aspartate aminotransferase 57 IU/L, alanine aminotransferase 48 IU/L, lactate dehydrogenase 1175 IU/L, and haptoglobin < 10 mg/dL. Originally, patient was diagnosed with HELLP syndrome. However, due to minimal elevation of AST, an alternative diagnosis was sought. Peripheral blood smear was performed which revealed 2 schistocytes/hpf, which suggested TMA. Due to risk of TTP or HUS, patient was urgently transferred to a facility capable of plasmapheresis. After transfer, patient had improvement in condition with blood pressure control and work-up ruled out TTP and HUS. Patient was diagnosed with TMA secondary to hypertensive emergency and transferred back. On return, patient’s blood pressure was managed by multiple anti-hypertensives. Patient was started on hemodialysis for acute renal failure. Patient stabilized and was discharged to continue outpatient dialysis after 12 days.

Discussion: Hypertensive emergency can affect many different organ systems, causing complications such as retinal hemorrhages, pulmonary edema, and cerebrovascular hemorrhage. In pregnant patients, uncontrolled hypertension is even more dangerous as it may lead to fetal distress or demise. With the high prevalence of hypertension and poor prenatal care in this country, it is extremely important to be aware of possible rare manifestations of hypertensive emergency such as TMA in this case. Accurate diagnosis is vital as such patients will often improve with blood pressure control without need for plasmapheresis or other costly treatments. Accurate diagnosis and early intervention are extremely important to help prevent maternal or fetal mortality.
MULTICENTRIC CASTLEMAN DISEASE IN THE UNCONTROLLED DIABETIC

Authors: Jessica Y. Chambers MD, University of Texas at Austin Dell Department of Internal Medicine

Introduction: Castleman Disease is a lymphoproliferative disorder with an array of well-documented associations, most notably HIV and human herpesvirus 8 (HHV-8). It may present as unicentric or multicentric distribution of disease. Although rare, the mortality from untreated multicentric disease is high, especially in those with autoimmune dysregulation. The disease is often caught late in its progression.

Case Presentation: A 33 year old male with uncontrolled type II diabetes was admitted to the hospital one evening for an asthma exacerbation. While his respiratory status improved quickly with proper treatment, he asks why he has so many “lumps” all over his body. On questioning he admits he has lost 75 pounds in the past few months without trying, complains of significant night sweats, and notes he has not been treating his diabetes appropriately for over a year due to treatment fatigue. On exam he is cachectic, diaphoretic, and tachycardic with nontender lymphadenopathy of the anterior and posterior cervical chains, as well as the axillary, epitrochlear, and femoral regions bilaterally. Laboratory studies were notable for a moderately elevated ESR and LDH, as well as a fasting blood glucose level of over 600 mg/dL without ketones in the serum or urine. A1C level was greater than 14. HIV testing was negative. Serum protein electrophoresis showed a faint monoclonal protein in the alpha-2 region and a polyclonal increase in gamma globulins. Imaging revealed diffusely prominent intrathoracic and inguinal lymph nodes over 1 centimeter in diameter. Excisional biopsy of the left inguinal lymph node exhibited follicles with atretic germinal centers containing prominent hyalinized vessels. Immunohistochemistry was negative for HHV-8. Interleukin-6 (IL-6) levels by multiplex bead assay were undetectable.

The patient was in the hospital for one week, during which time his glucose was aggressively controlled. His lymphadenopathy objectively decreased in size, and his fatigue was resolved. When seen in primary care clinic months later, he had self-discontinued all diabetic medications, including insulin. His disease appeared to have progressed, and his malaise had worsened. He reluctantly agreed to restart his medication regimen at a less aggressive level. He is currently undergoing rituximab infusions under the direction of hematology-oncology.

Discussion: The salient point in this case is the onset of multicentric lymphoproliferative disease in the setting of significant glucose toxicity. Multicentric Castleman disease also has a well-described association with another constellation of symptoms called POEMS syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal gammopathy, and Skin or bone changes). It is probable this patient’s presentation is a variant of POEMS syndrome, with endocrinopathy manifesting as severe insulin resistance. Even more compelling is the evidence that glucose control may prevent the progression of disease in this patient. This adds to the growing body of literature suggesting autoimmune dysregulation as an important, and possibly unrecognized, complication of uncontrolled diabetes.
TEXAS CLINICAL VIGNETTE POSTER FINALIST - AHMED T CHATILA, MD

A Case of Pseudo-Foreign Body in the Esophagus

Authors: Ahmed Chatila MD, Ronald Samuel MD, Hamza Abdulla MD, Praveen Guturu MD

Introduction: Foreign body ingestion is a mainstay of medicine, but many times imaging may be deceiving and can lead to unnecessary interventions. This case demonstrates the importance of performing a thorough physical exam prior to subjecting patients to any subsequent tests or interventions in suspected foreign body ingestions.

Case Presentation: A 36-year-old male with past medical history of multiple foreign body ingestions and Impulse and Conduct Disorder presents with abdominal pain concerning for repeat ingestion of sharp foreign objects. Two days prior to admission, he admitted to swallowing a screw while asleep. Initial chest X-ray at an outside facility, showed a single screw in his esophagus. Patient was transferred to our facility and began to experience hematemesis with bright red blood, chest pain, odynophagia, and generalized non-radiating abdominal pain. Repeat imagining on admission revealed 2 retained screws, one located in the esophagus and another in lower gastrointestinal tract, following which a lateral chest X-ray was ordered to assess the status of the newly found screw. Lateral view imaging demonstrated that the newly discovered object was indeed on the surface of the patient’s chest, and was found taped chest on the x-ray table. Prior to imagining, a noted physical exam was noted to be unremarkable. Continued imaging showed that there were still previously ingested screws in the small bowel. Patient was started on pantoprazole 40 mg IV BID, and serial imaging was ordered to assess eventual progression of the objects through the GI system.

Discussion: Inadequate and incomplete physical exams are often overlooked causes of medical errors and are not reported frequently in literature. These errors can lead to unnecessary testing and interventions, a potentially harmful hospital course, and delays in diagnosis or treatment. This result is preventable with further emphasis on thorough physical exams. A cross-sectional study looking at medical errors reported that 63% of errors were due to a failure to perform a physical exam, 14% of reported errors were due to a misinterpretation of the correct physical exam maneuver, and 11% showed a complete miss or failure to look for correct signs. These errors all led to diagnostic and treatment delays/misses, unnecessary healthcare costs, and many times preventable exposure to radiation or contrast [1]. In respect to our patient, a thorough exam prior to the chest X-ray would have revealed that the foreign body was simply taped to the abdomen rather than inside the patient’s distal esophagus, sparing the patient a lateral view x-ray. This case portrays a unique example of preventable error in medicine, that may have led to an unnecessary EGD on a patient. We would also like to remind all physicians of the importance of complete and correct physical exam maneuvers when assessing a patient prior to imagining and planned interventions.

References

Infectious Masses Mimicking Cancer

Authors: Alejandro Granillo, MD (ACP Resident); Siraya Jaijakul, MD, FACP (ACP Member)

Introduction: Coccidioidomycosis is an endemic and relatively common infection in the dry regions of the Southwestern United States. However, only about 1% of infected persons, with immunocompromised patients being more commonly affected,1 develop disseminated disease.2 In cases of suspected disseminated coccidioidomycosis, malignancy should always remain an important differential diagnosis.

Case Presentation: A 61-year-old Indian man with past medical history of type 2 diabetes mellitus and dyslipidemia presented with a 3-month history of intermittent dry cough associated with fevers, chills, night sweats, and generalized weakness. He worked in California as an engineer doing home remodeling, never smoked, and had no sick contacts or animal exposure. Upon admission, he was found to be septic and have acute hypoxic respiratory failure. Physical examination revealed temperature of 103.3°F Fahrenheit, heart rate of 107 beats/min, a respiratory rate of 35 breaths/min, and oxygen saturation of 87% on 32% fraction of inspired oxygen. Diffuse bilateral rales were present on lung auscultation. Pertinent laboratory findings revealed leukocytosis (WBC count 19.5k/µL) and elevated alkaline phosphatase of 325 U/L. Chest CT showed a left upper lobe subpleural mass with surrounding pulmonary infiltrate and scattered bilateral pulmonary nodules with associated lymphadenopathy. MRI of the abdomen showed multiple masses along the capsular surface of the right hepatic lobe and spleen. These findings were worrisome for malignancy. HIV Ag/Ab, T-Spot Tuberculosis, and Histoplasma antigen were negative. Serum Coccidioides IgM by ELISA was positive at 2.2 Index Value with a negative IgG. Coccidioides antibody by complement fixation was mildly elevated at 1:4. A bronchoscopy with transbronchial lung biopsy was performed, but failed to reveal evidence of malignancy; though both bronchial washing and tissue cultures grew hyaline mould, so a repeat bronchoscopy with biopsy was done. Pathology showed organizing pneumonia but bronchial washing culture eventually grew Coccidioides immitis. Therefore, the diagnosis of pulmonary coccidioidomycosis was confirmed. Unfortunately, the hepatic lesions could not be safely biopsied given their anatomical location so disseminated infection and malignancy cannot be entirely excluded. Patient was treated with intravenous liposomal amphotericin B and transitioned to high dose fluconazole with significant improvement of his symptoms and he would be followed up as an outpatient.

Discussion: Coccidioidomycosis was in fact the cause of this patient’s acute pulmonary findings, and it could possibly represent the hepatic and splenic masses. However, malignancy should always remain in a differential diagnosis in cases of suspected disseminated coccidioidomycosis as extrapulmonary coccidioidomycosis lesions are often indistinguishable from malignancy. Follow-up imaging studies of lesions should be performed. If there is no improvement in pulmonary and extrapulmonary lesions, further malignancy workup remains warranted. It is important to remember that one does not exclude the possibility of the other especially in a patient who is not known to be immunocompromised.

References

TEXAS CLINICAL VIGNETTE POSTER FINALIST - REX S HUANG, MD

(Sporo)Thrix or Treat: A rare case of disseminated Sporotrichosis

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Introduction: *Sporothrix schenckii* is classically known as the rose gardener’s thumb. This disease presents as small lesions that follow a linear progression up the forearm, following the lymphatic drainage and is usually easily treated with a moderate course of itraconazole. We present a rare and fatal case of disseminated sporotrichosis in an immunosuppressed patient.

Case Presentation: A 45-year-old gentleman native to the Rio Grande Valley, with Down’s syndrome, end stage renal disease on hemodialysis, and with a co-morbid history of repaired Tetralogy of Fallot was admitted for progressively worsening skin lesions over the last three months. On exam, BP 95/61, HR 98, T-max 99F, and RR 18 with SpO2 100% on 2L. There were multiple 1 cm x 1 cm erythematous painful skin lesions of the face and bilateral upper extremities in various stages of ulceration and pustular production. Vesicles were present, with oozing around the wounds. His family reported that he had recently been diagnosed with seronegative arthritis and had received two infusions of adalimumab, a TNF-α inhibitor.

Initial labs revealed a macrocytic anemia and elevated creatinine consistent with his end-stage renal disease. Blood, sputum, and urine cultures, Hepatitis panel, HIV, and TB-quantiferon were negative. Autoimmune workup was negative for Cyclic Citrullinated Peptide and Rheumatoid Factor. Biopsies were taken, which found granulomatous dermatitis, with an extensive differential of catastrophic antiphospholipid syndrome, ANCA vasculitis (primary or drug-induced), very rare reactions to TNF-α inhibitor (like adalimumab), or Ackerman's syndrome, an interstitial granulomatous dermatitis associated with autoimmune diseases.

He was treated with high-dose steroids, plasmapheresis, IV Immunoglobulins, as well as broad spectrum antibiotics (vancomycin and piperacillin-tazobactam) in addition to empiric antifungals (micafungin). Repeat skin biopsy from the left forearm showed epidermal denudation and underlying granulation tissue. PAS stain was negative for fungi. After 35 days, *Sporothrix schenckii* mold was isolated from the initial wound biopsies and our patient was diagnosed with disseminated sporotrichosis in the setting of his immunosuppressed state.

Discussion: Our case illustrates that *Sporothrix schenckii* is never a contaminant when found on biopsy. It is a dimorphic fungus that is inoculated by soil, plants, or other organic matter via trauma. It is typically treated with itraconazole, however in the Rio Grande Valley, Sporothrix is known to have resistance to this medication, potentially requiring amphotericin B in addition to optimization of wound care. In our patient, we recognized that he spent hours playing with small potted plants at bedside brought in by his family. Sporothrix can be found in the dead sphagnum moss in the soil of these plants, and we believe this is most likely how our patient came in contact with the fungus. Although we pursued the appropriate workup and treated our patient quite broadly, he eventually succumbed to his disease.

References

Cerebral vein thrombosis – a rare culprit of newly diagnosed seizure

Authors: Galyna Ivashchuk, MD, Christine Loftis, BS, MS-III, Brandon Cantazaro, MD

Introduction: Central venous vein thrombosis is a rare etiology of stroke with a varied clinical spectrum ranging from visual problems to encephalopathy. Because of the various causes and nonspecific manifestations of the condition, it is important to keep it on the clinical differential when assessing a newly diagnosed seizure patient.

Case Presentation: A 45 year old Hispanic woman with past medical history of diabetes mellitus, hypertension, obstructive sleep apnea, morbid presented to the hospital for syncopal episodes and speech slurring. Stroke team was activated. CT head, CTA of the neck and CT brain with perfusion did not show any acute abnormalities. Patient was discharged home. She returned to the hospital with recurrence of generalized tonic-clonic seizures at home witnessed by family member. In the emergency department, patient was noted to have numerous other brief seizures lasting about 30 seconds. EEG performed was significant for focal encephalopathy in the right hemisphere. Due to ongoing seizures, levetiracetam was titrated up until no further seizures were noted. MRI of the brain was significant for right frontal pachymeningeal enhancement. Lumbar puncture was performed and was negative for infectious process. Blood serology negative for Brucella, Fungus, Histoplasma, TB, HIV. Patient underwent right cerebral angiogram which showed partially recanalized thrombosis of the right frontal vertical vein near the junction with the superior sagittal sinus. Hypercoagulability work-up was negative. Patient was discharged home on anticoagulation.

Discussion: New onset seizures are a common manifestation of central venous thrombosis. This is the presenting symptom in approximately 39% of cases. Other symptoms include headache, visual problems, focal motor deficits, and encephalopathy. Pathophysiology of the condition has two distinct mechanism. First of all, obstruction of venous structures decreases capillary perfusion pressure, increases venous pressures, and leads to blood-brain barrier disruption with resultant vasogenic edema and hemorrhage. Additionally, obstruction of the dural venous sinus leads to decreased absorption of cerebrospinal fluid and subsequent elevation of intracranial pressure. Central venous thrombosis commonly affects women with a median age of 34 years. Risk factors include prothrombotic states, inflammatory diseases, infection, head trauma, malignancy, hormonal therapy, pregnancy and puerperium. Patients with clinically suspected central venous thrombosis should be evaluated with brain imaging, as well as work-up for hypercoagulable states. Treatment is geared toward addressing the underlying cause, seizure prophylaxis, decreasing intracranial pressure, and anti-thrombotic therapy with either endovascular or pharmacological interventions. Early intervention is imperative for favorable treatment outcome in these patients. Our case is unique, is that no underlying etiology was found for this disorder. Our patient has remained seizure free on levetiracetam therapy.

References

TEXAS CLINICAL VIGNETTE POSTER FINALIST - GERARDO MEDEROS, MD

S1Q3T3 Leading to Early Suspicion of Pulmonary Embolism in Low-Risk Patient

Authors: Residents: Dr. Gerardo Mederos, Dr. Som Aftabizadeh, Dr. Prashanth Reddy, Dr. Douglas Johnson, Dr. Harsh Patel; Dr. Daniel Jipescu; Dr. Anand Subramanian; Dr. Aman Patel; Attendings: Dr. Senthil Thambidurai; Dr. Madhira Machaiah, MD; Dr. Iqbal Mughal, MD

Introduction: Acute pulmonary embolism (PE), while relatively common, may prove fatal without early suspicion and subsequent treatment. Many cases go undiagnosed, with young patients most at risk of being misdiagnosed as suspicion in this population is very low. Even with a variety of diagnostic modalities, a high clinical suspicion remains key for diagnosis. Here we present a case of a patient with no known risk factors whose electrocardiogram (EKG) findings led to an early investigation, diagnosis, and subsequent treatment of a massive pulmonary embolism.

Case Presentation: A 34 year old African American male with a past medical history of asthma presented to our emergency department with a chief complaint of non-specific chest pain with dyspnea. On arrival, the patient was hemodynamically stable. His heart rate was regular but tachycardic. CXR showed no acute process. His EKG showed sinus tachycardia with T-wave inversion in inferior and anterior precordial leads along with S wave in lead I and Q wave in lead III. The WELLS score for PE was calculated at 0 and PESI was Class I. Though not sensitive, the EKG findings increased our suspicion for PE. D-dimer was ordered and found to be elevated at 6,693. A stat computed tomography angiogram revealed a large saddle pulmonary emboli. LMWH therapy was initiated along with supportive care. Further work-up uncovered a diagnosis of hereditary antithrombin deficiency and he was referred for specialist follow-up.

Discussion: Acute pulmonary embolism in young males without risk factors is rare. The varying degree of clinical presentation makes diagnosing PE very difficult. Given the time sensitive nature of appropriate diagnosis and treatment of PE, it is important that health care providers recognize EKG findings characteristic of PE. These findings can incite suspicion in low risk patients and direct subsequent work-up and management in a timely fashion.

References

“Dangerous Switch”: Severe Hepatitis Due To Reactivation of Hepatitis B in Patient Coinfected with Human Immunodeficiency Virus Following Change In Antiretroviral Therapy

Authors: Inyang Ndebbio, MD, Krishna Kamineni, MD, Vignesh Kannan, MD, Igor Dumic, MD

Introduction: Hepatitis B Virus (HBV) and Human Immunodeficiency Virus (HIV) co-infection is relatively common, with HBV co-infection diagnosed in 6-14% of patients in developed countries. All patients diagnosed with HIV infection should be screened for HBV and Hepatitis C (HCV) infection. Patients with co-infection have worse prognosis and faster progression to cirrhosis. Diagnosing co-infection presents a unique treatment opportunity as some of the antiretroviral agents used in the treatment of HIV also have proven activity against HBV.

Case Presentation: A 50-year-old man with longstanding and well controlled HIV (undetectable viral load and CD4 count 800) and HBV coinfection was admitted to our hospital for several days of right upper quadrant pain, nausea, vomiting, dark colored urine and yellow discoloration of his skin and eyes. Three months prior to current presentation his primary HIV provider had switched his tenofovir/emtricitabine based regimen to dolutegravir/rilpavirine with undetectable viral levels. Neither patient nor his primary care physician was aware of his HBV status. He denied fever, chills, travel to hepatitis A or E endemic areas or exposure to animals or insects. Physical exam was notable for stable vital signs. He appeared chronically ill but was in no distress. There were no stigmata of chronic liver disease but he had icteric sclera and skin. Lungs were clear on auscultation, heart sounds regular, without murmurs and abdomen was soft without ascites or tenderness. There was no leg swelling or petechial rash. Laboratory findings were significant for hemoglobin of 10mg/dl and mean corpuscular volume of 104, Aspartate aminotransferase (AST) and alanine aminotransferase (ALT) elevation with peak levels on day 8 at 2867 and 3237u/L respectively. Total bilirubin level was elevated, eventually peaking at 20.4mg/dL. INR also peaked at 1.52. HBV DNA level was elevated to 530,000IU/mL. Despite a protracted clinical course, he thankfully did not develop acute liver failure as he never had encephalopathy. Following resumption of tenofovir/ emtricitabine regimen his symptoms abated, 14 days into therapy his HBV DNA level had significantly decreased to 1683IU/mL. The patient improved and was discharged to his primary care provider.

Discussion: HBV reactivation may be triggered by sudden alteration in immune function such as during chemotherapy, use of immunosuppressants for various autoimmune and inflammatory diseases as well as during medication withdrawal or adjustment. Our patient developed severe hepatitis due to inadvertent change in his antiretroviral regimen that no longer contained medication active against HBV. Our case exemplifies the importance of proper communication between patient and medical provider as well as among providers themselves. Additionally, this emphasizes the importance of regular testing for HAV, HBV and HCV immunity in patients with HIV infection. Guidelines regarding duration of antiviral therapy for HBV in patients who are co-infected with HIV are conflicted and unclear but majority of USA providers believe it should be continued indefinitely.
TEXAS CLINICAL VIGNETTE POSTER FINALIST - DHРUV RAJPUROHIT

Beware of the dog, it could cost you a leg.

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Introduction: Dog bites are known to occur in about 4.7 million Americans each year with more than half of those cases in children with a fatality rate of 20-30. The injuries can involve the superficial skin and soft tissue to severe life-threatening trauma involving skeletal structures and vasculature. Vascular injuries most commonly involve the upper extremities, head and neck region; injuries of lower extremities are extremely rare. Progressive claudication with complete arterial occlusion post canine bite is extremely rare and has not been widely reported in literature.

Case Presentation: A 25-year-old Caucasian male with a past medical history of asthma and tobacco use presented initially to the emergency department (ED) after a dog bite to his left leg. Patient was trying to break up a fight amongst his dogs and was bitten. Physical examination revealed intact pulses in bilateral lower extremities with 2cm puncture lacerations in the anterior shin and posterior leg. Initially, patient did not report significant pain, active bleeding or deep puncture wounds. Patient was discharged home from ED on oral antibiotics. He did not seek medical attention for over 8 months despite noting subtle symptoms of pain, leg cramps, and numbness. As patient started to develop persistent toe numbness, calf tightness with leg pain on exertion and rest, along with reduced bulk and tone, he decided to pursue medical care. Patient was then referred to a cardiologist. Patient was started on dual antiplatelet agents Aspirin, Plavix, and Cilostazol along with a surgical consultation. Without significant relief post conservative management, the patient was then evaluated for surgical options due to the concern for impending critical limb ischemia. A CT Angiogram of the left lower extremity showed complete occlusion of the distal third of the superficial femoral artery (SFA), anterior tibial artery, popliteal artery, and peroneal artery with evidence of small collateral flow. Patient underwent successful percutaneous intervention with laser atherectomy, balloon angioplasty, and stenting of distal SFA.

Discussion: While dog bites are extremely common, arterial complications requiring surgical interventions are extremely rare. Most common type of vascular trauma post a canine bite is occlusion. Chronic limb ischemia occurs in around 10 million Americans and of those about 150 patients progress towards critical limb ischemia. It is of vital importance that physicians conduct a thorough history and physical examination to establish and correlate symptoms chronologically to unmask the underlying threatening diagnosis of critical limb ischemia. Our case highlights two distinct, however very important factors: recognizing the spectrum of injuries related to canine bites as well as early recognition of the progression of chronic limb ischemia towards critical, acute limb ischemia to allow for revascularization techniques to salvage the affected limb.
Severe Vitamin B12 Deficiency presenting as Hemolytic Anemia and Pancytopenia

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Introduction: Vitamin B12 deficiency is present in an estimated 6% of patients older than 60 years in the United States (1). However, Vitamin B12 (Cobalamin) deficiency as a cause for hemolytic anemia is rather unusual (2). Here we present a rare case of severe Vitamin B12 deficiency with hemolytic anemia and pancytopenia.

Case Presentation: A 63-year-old woman with no previous history and lack of medical attention over three decades, reported severe shortness of breath with generalized weakness and fatigue for two weeks. She reported an unstable gait, dizziness, two falls within the last year and memory loss. Her diet consisted of vegetables and meats; however, she had an unintentional two-pound weight loss over two months with poor oral intake. She denied hematochezia, hematemesis or melena.

On exam, patient had bilateral temporal wasting and pale conjunctiva. She had angular cheilitis and extremely poor dentition with multiple discolored, cracked and missing teeth. On neurological exam, she had 5/5 strength in all extremities and no difficulties with proprioception, pinprick or fine touch, or cerebellar disfunction, but had difficulty walking on her heels.

Admission labs noted pancytopenia with WBC 4.5 th/uL, Hemoglobin 5.1 gm/dL, MCV 125.9 fl and RDW 18.3 %, Platelets 91 th/uL, and Immature reticulocyte fraction 18.4%. She was transfused two units of packed RBCs with improvement of symptoms. Fecal occult blood test was negative and peripheral smear demonstrated pancytopenia with occasional oval macrocytes, increased polychromasia, and coarse basophilic stippling. Vitamin B12 level 53 pg/mL, Folate of 20.1 ng/mL, lactate dehydrogenase (LDH) 1706 IU/L, and Haptoglobin < 15 mg/dL. Iron level 241 ug/dL, Saturation 90%, TIBC 267, and Ferritin 145.3 ng/mL. Autoimmune workup confirmed elevated anti-parietal cell antibodies at 43.7 units with negative anti-intrinsic antibodies. She began 2000 mcg of daily oral Vitamin B12 supplementation, with clinical improvement after six weeks and complete resolution of neurological symptoms at 6 months. She will require oral maintenance therapy of Vitamin B12 at 1000 mcg for the remainder of her lifetime.

Discussion: Although uncommon, it is imperative that malabsorptive Vitamin B12 deficiency is recognized and treated because it is a reversible cause of bone marrow failure and demyelinating nervous system disease (3). Cobalamin is essential for DNA synthesis and hematopoiesis, and a severe deficiency results in megaloblastic anemia and pancytopenia secondary to the hypercellular and dysplastic bone marrow (4). This ineffective erythropoiesis and subsequent intramedullary hemolysis led to the elevated LDH. The combination of elevated LDH and low haptoglobin are 90% specific for diagnosing hemolysis (5). Cobalamin stores take several years before depletion secondary to malabsorption. Our patient’s lack to follow up with her doctor contributed to her undetected hemolytic anemia caused by her anti-parietal cell antibodies and complicated by her poor dietary intake.

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TEXAS CLINICAL VIGNETTE POSTER FINALIST - LILIAN VARGAS, MD

A Not So Typical Case of a Red eye

Authors: Lilian Vargas, MD, Hilda Mariana Gonzalez, MD, Monisha Singh, MD

Introduction: Red eye and eye pain are two common symptoms that can be associated to multiple diagnoses ranging from benign, self-limited conditions to significant systemic illnesses. Lung cancer typically presents with respiratory or constitutional symptoms, however symptoms associated with distant metastasis can be the first indicators of the disease.

Case Presentation: A 52-year-old man with no significant past medical history presented with right eye pain and redness for five days. He first noticed right eye redness and headache one day after snorkeling in Grand Cayman. He denied changes in his vision, discharge, fever, chills or rashes. He also denied cough, dyspnea, hemoptysis, fatigue or weight loss. He was a life-time non-smoker and had worked in Iraq and Afghanistan for five years for military purposes. Physical examination was unremarkable except for mild conjunctival injection of the right eye. Retinal examination revealed multiple bilateral amelanotic choroidal masses with the largest tumor involving the right macula. The lesions were concerning for metastatic disease. Of note, he had a normal eye examination one year before presentation. Laboratory studies showed an elevated carcinoembryonic antigen (229 ng/mL) and mildly elevated alkaline phosphatase (165 U/L). TB-Quantiferon was negative. Patient underwent computerized tomography of neck, chest, abdomen and pelvis that showed multicompartmental mediastinal lymphadenopathy, a moderate left pleural effusion, a small pericardial effusion and multiple lytic lesions in ribs, vertebral bodies, scapula and sternum. Left pleural effusion cytology and a supraclavicular lymph node biopsy demonstrated adenocarcinoma positive for CK7 and TTF-1, and negative for CK20 and CDX-2 which favored a lung primary. The patient was diagnosed with stage IV lung adenocarcinoma. Molecular testing revealed an EML4-ALK1 translocation. His course was complicated by the development of cardiac tamponade for which he underwent pericardial window. Pericardial biopsy and fluid cytology were also positive for adenocarcinoma. The patient was started on alectinib, an anaplastic lymphoma kinase (ALK) inhibitor. After eight weeks of treatment, there was resolution of eye pain and redness and improvement of choroidal masses in ophthalmological examination.

Discussion: This case illustrates an atypical presentation of lung cancer. In non-smokers, other risk factors for the disease should be explored. Asbestos, radon, ionizing radiation, and polycyclic aromatic hydrocarbons are known carcinogens for lung cancer. The patient possibly had prior occupational exposure to hazardous substances. Mild ocular symptoms associated with choroidal metastases are an uncommon presentation of malignancy, however it has been described in lung and breast cancer. Prompt and comprehensive work-up for identification of an underlying primary malignancy is essential for treatment. Fortunately for our patient, the identification of an ALK gene rearrangement allowed the use of targeted therapy for his adenocarcinoma with excellent results.

References
Blues Clues: Identifying Acquired Methemoglobinemia Early in the Acute Hospital Setting

Authors: Kevin Y. Wu, Rogerio Montes, Supraja Thunuguntla, Joel J. Pallapati, Javier A. Cabello Garza

Introduction: Acquired methemoglobinemia (AMetHb) after administration of local anesthetics for transesophageal echocardiogram (TEE) procedures is an uncommon, yet dangerous, occurrence where rapid recognition offers readily available solutions and avoids the costly workup. Here we describe 2 cases where prompt awareness can guide the hospitalist to the correct diagnosis and treatment.

Case Presentation: A 79-year-old male presented to the hospital with gross hematuria and flank pain after ureteral stent placement for bladder neck contracture. After stent removal, the patient developed fevers and was found to have MRSA bacteremia. TEE was ordered and aborted shortly when patient desaturated to 79% and was cyanotic after benzocaine spray given for local anesthetics. The patient was placed on 100% venturi mask and then transferred to the intensive care unit for further management. PaO2 on arterial blood gas (ABG) was >400, pulse oximetry was 88%, but the patient was not in respiratory distress. With high suspicion for methemoglobin (MetHb) toxicity related to anesthetic spray, intravenous (IV) methylene blue and vitamin C was given before MetHb levels returned. Within 30 minutes, O2 saturation improved on room air and the patient was transferred to the ward the next day. MetHb levels measured by co-oximetry resulted at 26%. In another similar case, a 71-year-old male undergoing stroke workup with TEE had O2 desaturation to 80% and central cyanosis afterward. Once again, MetHb toxicity was suspected and intravenous methylene blue was given without vitamin C and patient’s O2 saturation improved. MetHb level was elevated at 41%. Upon further questioning, it was discovered that in addition to the liberal 2-second benzocaine spray that was given before the TEE, he was also given a swish and spit of 2% viscous lidocaine. In both cases, there were no presentations of acute distress and a dark chocolate brown ABG was pathognomonic.

Discussion: AMetHb associated with TEE can be fatal if missed. When the clinical picture arises with strong evidence of low O2 saturation, normal ABG values, chocolate-brown arterial blood in a patient without severe distress, then MetHb toxicity should be prioritized. And while waiting for the cost-effective treatment of methylene blue or vitamin C to take effect, usually within an hour, the clinician can consider other deadly complications from the TEE. There is also no objective measure to determine the amount of local anesthetic spray that is given before any TEE, so this will be a quality measure that will need further dissection. Both cases above presents opportunities for the clinician to astutely identify the issue within the context of the patient’s hospital course and address the complication without costly extensive workup.
An unusual cause of bowel obstruction: colonic mucormycosis

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Introduction: Mucormycosis is a rare but serious fungal infection caused by environmentally ubiquitous mucormycetes. While it is classically associated with infections of the head, sinuses, orbits and cerebrum, it can affect any part of the body. Infection occurs primarily in immunocompromised and/or diabetic patients. We report a case of colonic mucormycosis in an immunocompromised patient.

Case Presentation: Our patient is a sixty-two year old female with myelodysplastic syndrome that converted to acute myeloid leukemia who was admitted and underwent haploidentical stem cell transplantation (HSCT). During the month prior to admission, she had neutropenic fever with enteritis/colitis for which she underwent sigmoidoscopy with biopsy which did not show fungal elements at that time. Following readmission for HSCT, her hospital course was prolonged due to delayed engraftment. Within a few days post-transplant, she started experiencing abdominal pain and decreased stool output. Abdominal computed tomography (CT) demonstrated a 4.9 cm perirectal abscess. Cytology of the abscess aspirate revealed aseptate fungal hyphae concerning for mucormycosis. (1,3)-β-D-glucan and galactomannan assays were negative, and she had been taking posaconazole for several months prior to HSCT. Due to concern for mucormycosis, CT imaging of the sinuses, head and chest was obtained and was negative for other foci of infection. Liposomal amphotericin B and micafungin were initiated and posaconazole was stopped. Fungal cultures from the abscess grew Rhizopus arrhizus. The patient underwent a proctosigmoidectomy with colostomy to remove the fungal abscess. However, six days after her surgery the patient had return of fevers and abdominal pain. Repeat abdominal CT revealed recurrence of the fungal abscess despite recent surgical excision and antifungals. Given the high likelihood of poor surgical outcome for repeat debridement/pelvic evisceration, the patient declined further interventions. She was discharged to hospice with palliative/salvage isavuconazole. However, the patient remained clinically stable and repeat imaging one month following discharge revealed interval improvement of the abscess without complete resolution.

Discussion: Mucormycosis is a feared fungal infection in the immunocompromised or diabetic patient with high morbidity and mortality. While rhino-cerebro-orbital infections are classically associated with mucormycosis, atypical foci of infection may occur as our case demonstrates. While previously more frequently described in neonates and infants with gastric infection or necrotizing enterocolitis, gastrointestinal mucormycosis is being described more recently in iatrogenically immunocompromised patients. Route of entry is assumed to be ingestion. Despite surgery and antifungal therapy, mortality remains high as does subsequent dissemination. Our case represents a rarity in its presentation and clinical course. With recent sigmoidoscopy and biopsy in the preceding month, post-procedural infection may have led to her presentation. Our case also potentially supports isavuconazole for salvage therapy following debridement in mucormycosis.
A Rare Potential Mimicker of Autoimmune Encephalitis

Authors: Kevin J. O’Gorman, MD; Timothy R. Fullam, MD; Thomas M. Duginski, MD; John HWA Sladky, MD

Introduction: Advances in neuroimmunology over the past decade have led to increased recognition and empiric treatment of presumed autoimmune encephalitis. Despite these advances, there remain well described, but rare, clinical entities which may mimic antibody negative autoimmune encephalitis.

Case Presentation: 25-year-old female with history of migraine headaches initially presented following a generalized seizure at home. Initial workup to include laboratory studies and CT Head were negative and the patient was started on empiric anti-epileptic drug treatment. Electroencephalography (EEG) obtained as an outpatient was remarkable for symmetrical background slowing. MRI Brain was significant for left pre and post central gyrus edema, restricted diffusion with mild pachymeningeal enhancement, and bilateral putaminal T2 hyperintensities. Three months later, the patient was hospitalized for status epilepticus and imaging obtained during admission demonstrated persistent bilateral putaminal hyperintensities, new right parafalcine parietal lobe hyperintensities, and interval resolution of the left parietal lobe findings noted previously. A diagnosis of autoimmune encephalitis was made despite normal CSF studies and multiple negative serologies, and the patient was treated with a combination of intravenous solumedrol and IVIG. One year after the patient’s initial presentation, the patient was transferred to our facility after arriving to an outside hospital in status epilepticus. Laboratory workup to include CSF studies and repeat serology testing were unremarkable. EEG performed at this time was consistent with a diffuse encephalopathy without evidence of ongoing status epilepticus. MRI demonstrated restricted diffusion and faint enhancement in the left occipital lobe as well as continued hyperintensity of the bilateral putamen. MRI Spectroscopy demonstrated an elevated lactate peak in normal appearing grey matter. Given the combination of an unexplained seizure disorder and encephalopathy, stroke like lesions at a young age, bilateral putaminal lesions, elevated lactate peak, and previous normal development, there was high clinical suspicion for Mitochondrial Myopathy, Encephalopathy, Lactic Acidosis, and Stroke-like episodes (MELAS). Initial MELAS genetic panels were negative and a muscle biopsy was equivocal. Follow up whole exome sequencing and mitochondrial DNA analysis revealed a pathogenic MT-ND5 gene mutation in the mitochondrial DNA which confirmed the diagnosis of MELAS.

Discussion: While rare, MELAS can mimic the presentation of autoimmune encephalitis in young adults because seizures and altered mentation are common initial manifestations of the disease. It should remain on the differential in individuals with migrating, stroke-like lesions on advanced neuroimaging, persistent encephalopathy despite immunotherapy, and otherwise negative workup for autoimmune or paraneoplastic etiologies. Roughly 80% of cases of MELAS are associated with the m.3243A>G mutation in the MT-TL1 gene with other pathogenic mutations in this gene accounting for another 15-20% of cases. Rarely, mitochondrial gene mutations may be the underlying cause. Less than ten MELAS cases with an MT-ND5 pathogenic mitochondrial gene mutation have been reported in the literature with the above case representing an additional case of MELAS with this particular gene mutation.

References


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Mycobacterium Tuberculosis Meningoencephalomyelitis Induced Optic Nerve Atrophy and Permanent Blindness

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Introduction: It is estimated one-third of the world population are infected with Mycobacterium Tuberculosis (MTB). Central nervous system disease accounts for less than 1% of all cases in the developed world and typically occurs in the reactivated state. Rarely before has it been documented to induce irreversible, progressive neurologic deficits despite appropriate treatment as its initial presentation.

Case Presentation: The patient is a 21 year old male with no past medical history who presented with leg weakness and altered mental status. Magnetic resonance imaging (MRI) revealed multiple enhancing cortical and subcortical lesions without mass effect. Chest computed tomography revealed bilateral upper lobe reticular infiltrates. Lumbar puncture found pleocytosis and negative gram stain with polymerase chain reaction finding pan-susceptible MTB. Successive sputum samples cultured MTB. He was started on rifampin, isoniazide, pyrazinamide and ethambutol (RIPE) for MTB meningoencephalomyelitis and discharged in a stable condition. He re-presented two weeks later with fevers, somnolence, dysmetria, bilateral cranial nerve six and left sided cranial nerve seven palsies. He received IV antibiotics, steroids, and underwent lumbar puncture with unchanged findings. He developed urinary retention and hyponatremia. With no symptomatic resolution over the following days, repeat imaging revealed obstructive hydrocephalus with unchanged cerebral lesions. He underwent external ventricular drain with marked neurologic improvement albeit continued debilitating blurry vision. He discharged to the Texas Center for Infectious Disease (TCID) with the addition of levofloxacin to his RIPE therapy while he received high dose Rifampin 1500mg daily. With a prolonged steroid taper he gained 90 pounds over the next 10 months. Subsequent MRI imaging revealed resolution of multiple cerebral lesions. However, he had bilateral optic nerve atrophy and progressive growth of a left frontal lobe mass. He underwent surgical resection. Pathologic specimens showed caseating granulomatous inflammation with positive acid-fast smear and negative cultures consistent with dead MTB. He returned to the TCID for completion of his treatment, walking out of the hospital near his original weight upon discharge, legally blind, cured of MTB.

Discussion: This case highlights an extreme presentation of a pan-susceptible MTB pulmonary and meningoencephalomyelitis infection in an immunocompetent patient causing bilateral optic nerve atrophy and permanent blindness, a seldom before documented complication of MTB. Although he developed debilitating lower extremity weakness and urinary retention, he had near full neurologic recovery with antibiotic and steroid therapy. While steroids are imperative in the treatment of MTB meningoencephalomyelitis, it was at the expense of extreme weight gain complicated by a singular unchecked tuberculoma. MTB therapy has remained largely unchanged over the last few decades, with MTB meningitis requiring high-dose Rifampin. Commonly treated for 9-12 months for pan-susceptible infection, this patient afflicted with delirium, blindness, cranial nerve palsies and paraparesis underwent 24 months of therapy with a near full recovery.

References

3. 

A Rare Case of Epithelioid Sarcoma

Authors: Amy Stacey, DO, (ACP Member), Amanda Bilko, MD, Divya Indrakanti, Thomas Murphy, MD.

Introduction: We present an unusual presentation of an aggressive chest wall epithelioid sarcoma in a young adult male with normal chest radiograph two months prior.

Case Presentation: A 27 year-old male presented with two months of persistent left-sided chest and shoulder pain. Prior emergency department workup was unremarkable, including a normal chest x-ray (Figure A). Patient reported subjective fever, chills, night sweats and periodic hemoptysis for two weeks. He had no contributable social, family or travel history. On exam, he was tachycardic (104 bpm) and febrile (38.2°C). Labs demonstrated a leukocytosis (17.2k). Chest x-ray showed a new large left apical mass (8 cm) when compared to imaging two months earlier (Figure B). CT chest revealed a pleural-based lesion in the left upper thorax with extra-pleural invasion involving the fat and third rib (Figure C, D). Broad spectrum antibiotics were initiated. Bronchoscopy failed to identify source of bleeding or abnormal tissue for biopsy. Preliminary pathology from CT-guided biopsy revealed poorly differentiated malignant cells. Thoracotomy with wedge resection was attempted and then aborted due to near total left upper lobe encasement, chest wall invasion and diaphragm metastases. Empiric chemotherapy was started for presumed small cell lung cancer. The patient received three cycles of carboplatin and etoposide with minimal tumor regression. Final pathology verified the absence of INI-1, confirming the diagnosis of metastatic INI-1 deleted undifferentiated pleomorphic sarcoma, likely epithelioid sarcoma. The patient was started on AIM therapy (Adriamycin, Ifosfamide and Mesna) and was referred to a Sarcoma Alliance cancer center.

Discussion: Epithelioid sarcomas (ES) account for <1% of adult malignant tumors. ES occur in young to middle-aged males and arise in the head, neck, trunk, pelvis and perineum. Core needle biopsy demonstrates epithelioid cells with large vesicular nuclei in a sheet-like pattern. About 90% of ES have an inactivation of INI-1, a tumor suppressor gene. Treatment involves radical resection with adjuvant chemotherapy and radiotherapy. Early metastases and a large primary unresectable tumor have a poor prognosis. Recurrence rates and metastatic potential are very high. Overall five-year survival is 50%. This aggressive presentation initially triggered infectious management; however, histology confirmed a rare undifferentiated pleomorphic sarcoma consistent with epithelioid sarcoma. Even more unusual was the tumor's involvement of the chest wall. A literature review revealed six prior cases of chest wall ES, including this case. This presentation of rapidly progressive epithelioid sarcoma, discovered in a previously healthy young male, highlights a rare but significant diagnosis that unfortunately carries a poor prognosis.
Type 5: A Rare MODY Operandi

Authors: Zachary Bloomer, MD, Roy E. Guinto, DO, FACP, Ian Rivera, MD, FASN

Introduction: A case presentation and review of maturity onset diabetes of the young type 5, in a 33 year-old man with renal cysts and pancreatic atrophy with no previous family history.

Case Presentation: A 33-year-old man with a history of diabetes and renal insufficiency presented to the endocrinology clinic for management of his diabetes and pancreatic atrophy noted incidentally on computed tomography scan. His diabetes was not controlled on oral medications since his diagnosis eight years prior to presentation. Initial diabetic evaluation made both autoimmune destruction of the pancreatic beta cells and peripheral insulin resistance unlikely. Genetic analysis revealed a mutation of the hepatocyte nuclear factor-1β gene confirming the syndrome previously known as maturity onset diabetes of the young type 5 (MODY5). This condition is a rare diagnosis representing about 1-5% of diabetics. It occurs via genetic mutation resulting in beta cell dysfunction and decreased insulin secretion.

Discussion: MODY5 is a rare genetic disease resulting in decreased insulin secretion presenting as non-ketotic diabetes prior to age 25 with pancreatic and renal abnormalities. Recognizing the features of atypical diabetic presentations can lead to earlier diagnosis, improved education, counseling and management for patients and their families.
Lessons in lumbago: an acute presentation of Guillain-Barre Syndrome

Authors: CPT Nicholas Hodgeman, Capt Lacy Lowry, Lt Col Sky Graybill

Introduction: Lower back pain is one of the most common patient complaints and less than 1% have serious systemic etiologies. Most clinicians are trained to assess for cauda equina syndrome, but other important causes are often forgotten, particularly those that do not show abnormalities on anatomic imaging. Guillain-Barre Syndrome (GBS) is one of these conditions; it is notorious for ascending paralysis but lumbago is present in a surprisingly high number of these patients (66%). Symptom onset is typically 1-3 weeks following an upper respiratory or gastrointestinal illness, and consists of rapidly progressive ascending areflexic motor possible. The rapid and serious nature of the disease coupled with the often benign presentation makes the diagnosis challenging.

Case Presentation: A 57-year-old man presented with one week of sharp lower back pain that radiated into his legs. He also had fatigue, a 10-pound weight loss, as well as finger and lip paresthesias. He had an upper respiratory infection several week prior. He denied fever, night sweats, insect bites, rash, saddle anesthesia, and loss of bladder or bowel function. He had visited local urgent care centers twice in the preceding week for lumbago for which he received analgesics and muscle relaxants. Physical exam was notable for preserved sensation in the lower extremities and orthostatic hypotension responsive to fluids. Complete blood count, complete metabolic profile, creatine kinase, thyroid stimulating hormone, urinalysis, and lumbar spine computed tomography were unremarkable.

He was subsequently admitted to evaluate for a possible spinal infection. However, magnetic resonance imaging of the brain and spine was unremarkable. He developed a unilateral facial droop, dysphasia, and lower extremity weakness. Lumbar puncture then demonstrated albuminoocytologic dissociation consistent with GBS. He received intravenous immunoglobulin (IVIg) and had improvement in motor function.

Discussion: This case highlights challenges with managing lumbago. GBS can present with back pain but normal imaging, leading to false reassurance. Diagnosis requires a high index of suspicion because prompt treatment with IVIg or plasmapheresis before the disease plateaus improves prognosis.
Rapidly Progressive Interstitial Lung Disease in a Patient with a Rare Antisynthetase Antibody and Concomitant Anti-Ro52 Positivity

Introduction: Antisynthetase syndrome is a rare, autoimmune disease spectrum characterized by myositis, arthritis, interstitial lung disease (ILD), Raynaud’s phenomenon, and distinctive cutaneous manifestations. The serologic hallmark of the disease is the presence of myositis specific autoantibodies (MSA), of which anti-Jo1 is the most common. These autoantibodies are specific for the condition, have historically been considered mutually exclusive, and appear to predict a distinct clinical phenotype. Recent literature has shown that anti-Ro52, a myositis associated antibody (MAA), predicts an increased incidence of ILD and myositis when found concomitantly with anti-Jo1. Anti-PL12 is a rare MSA that has been associated with an amyopathic presentation and rapidly progressive ILD. There is a paucity of literature describing the phenotypic expression, disease course, and response to treatment associated with co-positivity for anti-PL12 and anti-Ro52. We describe a case of a patient with these two autoantibodies who presented with rapidly progressive ILD and myositis ultimately requiring two steroid pulses and intravenous immune globulin (IVIG).

Case Presentation: A previously healthy 58-year-old man developed rapidly progressive respiratory distress requiring supplemental oxygen and monitoring in the intensive care unit (ICU). Onset was associated with bilateral thigh pain and hyperkeratosis of hands and feet consistent with mechanic’s hands and hiker’s feet. Laboratory results were notable for anti-PL12 by immunoprecipitation and anti-Ro52 by enzyme-linked immunosorbent assay (ELISA). Creatine kinase was elevated to 1031 U/L. EMG was consistent with a myopathic process, and he was found to have nonspecific interstitial pneumonia (NSIP) on chest computed tomography (CT). Pulmonary function testing revealed moderately severe restriction with decreased diffusion capacity of the lungs for carbon monoxide (DLCO). His lung disease responded well to methylprednisolone pulse. However, he developed rapidly progressive proximal weakness and symptoms concerning for early dysphagia with rise in creatinine kinase to 1394 U/L while on high dose prednisone. He was treated with a dexamethasone pulse and IVIG with excellent improvement in the myopathy.

Discussion: Literature suggests that myositis specific autoantibodies can predict a distinct clinical phenotype within the antisynthetase syndrome spectrum. When compared to the more common anti-Jo1 phenotype, patients with anti-PL12 antibodies have lower rates of myositis and higher rates of ILD. There is a paucity of literature describing the clinical phenotype profile of concomitant anti-PL12 and anti-Ro52 positivity. This patient’s presentation suggests that the presence of anti-Ro52 is associated with rapidly progressive lung disease and myositis in anti-PL12 positive patients. After initial disease flare, our patient responded well to dexamethasone and IVIG. Co-positivity of these two antibodies should raise concern for propensity of respiratory decompensation and significant myopathy. Further descriptive studies are necessary to determine the exact clinical phenotype and response to treatment in this rare patient population.

References


US ARMY CLINICAL VIGNETTE POSTER FINALIST - CAROLINE MURPHY

Blue Lips n’ Fingertips: Positional Cyanosis Reveals Platypnea-Orthodeoxia-Syndrome

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Introduction: Platypnea-orthodeoxia syndrome (POS) is a rare cause of dyspnea in which a sensation of breathlessness is caused by position-dependent hypoxemia; dyspnea is improved in the recumbent position and worsened with sitting or standing. POS is a rare and often unrecognized cause of hypoxemia, with only a few isolated case reports and case-series in the literature. The underlying cause of POS is felt to be either intra-cardiac shunting, pulmonary-vascular shunting, or ventilation-perfusion mismatches. Here we present a case of 79 year old female with who presented with worsening dyspnea accompanied by repeated cyanosis of her lips and fingertips, ongoing for months. Work-up revealed an atrial septal defect resulting in significant right-to-left shunt and significant hypoxemia while standing that resolved laying down. This case underscores the need for providers to consider platypnea-orthodeoxia in the differential in otherwise unexplained positional hypoxemia.

Case Presentation: A 79 year old female with a history significant for chronic obstruction pulmonary disease, coronary atherosclerosis, heart failure with reduced ejection fraction AHA stage C NYHA class II symptoms, and atrial fibrillation presented to her primary care physician complaining of acutely worsening shortness of breath. She was initially treated with optimization of her medical therapy as well as her cardiac medication regimen. Despite this, she noted that her lips and fingertips would occasionally turn blue, which in the context of continued dyspnea, raised concerns for ventricular unloading suggestive of a potential significant right-to-left intra-cardiac shunt. Review of her previous transthoracic echocardiograms and MRI suggested that she may have had an underlying secundum-type defect and she then underwent transesophageal echocardiogram (TEE) for further evaluation. The TEE confirmed findings of significant right ventricular systolic dysfunction and dilatation as well as significant right atrial chamber enlargement. Evaluation of her atrial septum revealed evidence of what appears to be a moderate size patent foramen ovale with evidence of right-to-left shunting by agitated saline at rest. A right heart catheterization revealed upper limit of normal filling pressures and upper normal pulmonary. The patient was found to have a resting oxygen of 95% throughout the case compared with 89% while standing. The patient was diagnosed with platypnea-orthodeoxia syndrome and prescibred home oxygen which adversely affected her quality of life, thus the patient will undergo definitive closure.

Discussion: Platypnea-orthodeoxia syndrome (POS) is an uncommon condition of positional dyspnea (platypnea) and hypoxemia (orthodeoxia). The symptoms occur when the patient is upright and resolve quickly with recumbency. These findings are the opposite of those typically seen in cases of advanced heart failure and can pose a diagnostic dilemma. Platypnea-orthodeoxia syndrome should be suspected when an individual has normal oxygen saturations on room air while supine, yet experiences dyspnea and desaturations.
Last Response for the First Responder?
Arrhythmogenic syncope in a soldier first-responder reveals rare channelopathy

Authors: Murphy, C.E, Fentanes, E, Department of Medicine, Cardiology Service, Tripler Army Medical Center

Introduction: Catecholaminergic-induced polymorphic tachycardia (CPVT) is a rare cardiac channelopathy, induced by stress or emotion, which affects approximately 1:10,000 individuals and is a cause of sudden cardiac death in persons with structurally normal hearts. The military population is unique in that candidates undergo medical screening and are disqualified for known cardiac disease; however, in autopsy analyses of military sudden cardiac death (SCD) events, channelopathies such as CPVT have been identified as the etiology in up to one-third of deaths. Here we present the case of an active duty first-responder with arrhythmogenic syncope induced by a sudden resuscitation effort, underscoring the need for providers to maintain clinical suspicion for channelopathies in patients with unexplained syncope, as well as to query the need for the military to perform genetic screening in individuals that work in stressful environments.

Case Presentation: A 26 year-old active duty nurse with a history of unexplained childhood syncope had a patient enter sudden cardiac arrest. While performing chest compressions the nurse syncopized, prompting activation of a rapid response team. Immediate evaluation, to include a twelve-lead EKG, was unremarkable and suggestive of vasovagal syncope. Further evaluation with cardiac MRI, transthoracic echocardiogram, and coronary CT angiogram revealed no abnormalities. A graded exercise tolerance test (GXT) was performed; it was observed that at heart rates above 125 bpm frequent PVCs developed in a bigeminy pattern and six minutes into her stress test nonsustained but bidirectional ventricular tachycardia was evident. Given the syncopal history and GXT findings, genetic testing was obtained that revealed heterozygosity in two variations in the RYR2 genes. Genetic findings in combination with clinical history were consistent with the diagnosis of CPVT.

Discussion: Catecholaminergic-induced polymorphic tachycardia is highly lethal and estimated to be the cause of 12% of unexplained sudden deaths in the general population. CPVT is caused by aberrant release of calcium leading to delayed afterdepolarizations resulting in ventricular tachycardia; this is exacerbated by adrenergic states associated with exercise, high-emotion or high-stress situations. Genetic testing may reveal mutations in calcium channels. Patients have normal cardiac findings but enter ventricular arrhythmias above 110bpm characterized as bidirectional; thus, stress testing is typically necessary. Prompt diagnosis is crucial, and management begins with beta blockers and limitations placed on activities to control heart rate. Implantable cardio-defibrillator therapy is recommended for anyone resuscitated from cardiac arrest, as well as those with syncope despite beta-blocker therapy or symptomatic patients with two or more known genetic mutations. CPVT must remain on the differential in individuals with recurrent syncope and structurally normal hearts, and evaluation must include exercise stress testing. Given the risk for high-stress work, the military may consider adding genetic testing to intake screening.
US NAVY CLINICAL VIGNETTE POSTER FINALIST - LCDR HILLARY A. CHACE, MC USN

Functional Neurologic Disorder: A must-know diagnosis for internists.

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Introduction: Functional neurologic disorders (FND), formerly known as psychogenic/conversion disorders, are clinical conditions defined by movements that are unusual and unrelated to a known neurological origin. According to the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5) diagnostic criteria require one or more symptoms of altered voluntary motor or sensory function with an incompatibility of symptoms with a recognized neurological condition, which are not better explained by another medical or mental disorder. Patients may present with symptoms such as blindness, paralysis, non-epileptic seizures, tremor or syncope and thus can be difficult in the outpatient setting to distinguish from a strictly neurological disorder.

Case Presentation: A 19 year old healthy male presented to the emergency department (ED) with 3 hours of muscle spasms in his left trapezius accompanied by persistent grunting, vocalizations and emesis, without incontinence or stupor. In the ED he experienced multiple convulsive episodes characterized by dystonic neck movements, bilateral upper extremity tonic posturing, rhythmic trunk movement, and tics with grunting, snorting, sneezing which abated after lorazepam and fosphenytoin. He maintained intra-event memory and had no postictal state. His labs were normal and advanced neuroimaging revealed no clinically significant findings. A continuous video electroencephalopathy (EEG) captured two spells consistent with prior reported episodes with normal background rhythm and no evidence of epileptiform activity. He was diagnosed with FND with dissociative spells, and was initially prescribed clonazepam with transition to escitalopram. The diagnosis was immediately disclosed to him, with initiation of multidisciplinary care with mental health and neurology. He is currently undergoing cognitive behavioral therapy with dramatic reduction in severity and frequency of attacks.

Discussion: This patient’s case is an example of FND manifesting as non-epileptic attacks. On first evaluation, it was thought that he was having epileptic seizures; however, positive features lending credibility to a FND include: long duration of spell, closed eyes during attack, memory of being in the seizure, ictal hyperventilation and normal ictal EEG. Patients often first present in the outpatient setting with an array of physical and psychosocial manifestations that can be overwhelming and seemingly unrelated. While the diagnosis of FND is made on positive findings, the possibility of co-diagnosis must be kept in mind. For seizures, an EEG capture of a typical event is the gold standard for diagnosis. In the outpatient setting a good approach to these patients is a systematic fashion by first focusing on detailed history and review of all organ systems including stress, fatigue, sleep, memory and concentration. Early diagnosis and prompt treatment offers the best prognosis for patients with FND.

References

Adjusting to Evolving Symptoms: Multiple Sites of GI Bleeding from the Same, Uncommon Cause

Authors: Matthew T Nelson MD, Aaron Tallant MD, Brett W Sadowski MD FACP, Joshua Davis MD, Anita Bhushan, MD

Introduction: Gastrointestinal bleeding is a common indication for admission to the hospital and typically be classified by either an upper or lower tract source by clinical and laboratory findings. However, depending on the etiology of the bleeding, presenting symptoms could vary and evolve, making diagnostic evaluations difficult. We present the case of an elderly male who had manifestations of both acute upper and lower GI hemorrhage eventually found to have a uncommon cause for both.

Case Presentation: An 81 year old man with a history of chronic idiopathic autoimmune neutropenia and a new month long history of progressive leukocytosis of unknown etiology presented with painless hematochezia for two days. He was initially admitted to the floor with plans to proceed with colonoscopy, but subsequently developed melena and hemodynamic compromise and was transferred to the intensive care unit for aggressive stabilization, requiring multiple units of packed red blood cells. He was eventually resuscitated, although he was once again having persistent hematochezia. He underwent a colonoscopy and upper endoscopy which demonstrated abnormal, erythematous, friable and ulcerated tissue in the terminal ileum, multiple clean based ulcers in the ascending colon and at the ileocecal valve, a large clean based ulcer in the rectum, as well as multiple erosions in the gastric antrum and duodenal inflammation in the bulb. Biopsies taken from each of these sites demonstrated both clonal B and T cell populations and diffuse plasma cells throughout the upper and lower GI tracts. After oncologic consultation and further testing, the diagnosis of aggressive B-cell plasmablastic lymphoma (PBL) as well as an indolent T-cell lymphoma was made. The patient was started on V-EPOCH (etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin, and bortezomib) based on therapeutic potential in previous case reports using this regimen for PBL. His hematological processes responded well to chemotherapy but after recovering from a long hospitalization, the decision was made to not proceed with further medical therapy.

Discussion: PBL is an aggressive lymphoma commonly associated with HIV or underlying immunodeficiency. GI involvement of PBL has been rarely described and can commonly present with bleeding as well as weight loss, anorexia and abdominal pain while potentially occurring at all sites in the luminal GI tract. This case is unique from most reports in the literature given the multifocality of the disease, which is consistent with the evolving symptoms with which the patient presented. The case highlights the need to consider a broad differential for common presentations and further underpins the importance of rapid diagnosis and interdisciplinary management.

References

When Latent Tuberculosis Stirs the Pott: A Case Study of Spinal TB

Authors: LT Melanie Wiseman, MD, Internal Medicine PGY1 WRNMMC (Associate), LCDR Christa Eickhoff, MD, Infectious Disease NMCP (Member)

Introduction: Tuberculosis (TB) is one of the world’s deadliest diseases; most commonly manifested as a pulmonary infection. [1] However, extra-pulmonary TB infection has become increasingly recognized in the United States. [2] This case report details a unique case of extrapulmonary TB with isoniazid resistance.

Case Presentation: A 62-year-old, otherwise healthy, African American woman reported to multiple emergency departments and her outpatient primary care provider several times over a span of approximately 2 months with subacute, worsening, atraumatic back pain without red flag symptoms. Magnetic resonance imaging (MRI) of the spine showed findings concerning for focal discitis and secondary osteomyelitis involving the 12th thoracic (T12) and 1st lumbar (L1) vertebral levels. She developed fevers and was started on empiric ceftriaxone and vancomycin for presumed bacterial vertebral osteomyelitis. A fourth generation HIV test was negative. However, she was originally from Jamaica and was previously a nurse, with a past medical history significant for untreated latent TB. Acid Fast Bacteria (AFB) stain of the histopathology specimen obtained via Computed Tomography-guided biopsy of the intervertebral disc was negative, but at 5 weeks Mycobacterium tuberculosis was detected by Gene Probe from the bone biopsy AFB culture. The patient was diagnosed with tuberculosis spondylitis, otherwise known as Pott’s Disease, and started on rifampin, isoniazid, pyrazinamide, and ethambutol (RIPE) for treatment. The TB strain was found to be susceptible to rifampin, but was resistant to isoniazid with katG mutation detected. At that time, the patient was transitioned to rifampin, pyrazinamide, ethambutol, and moxifloxacin. Her clinical course was complicated by right paraspinal abscesses that were drained, as well as multiple medication adverse effects. She developed drug-induced neutropenia from 4 months of treatment with rifampin 600mg daily, which resolved after cessation of rifampin and transition to bedaquiline, linezolid, moxifloxacin, pyrazinamide, and ethambutol. She subsequently developed drug-induced polyneuropathy from 3 months of linezolid 600mg daily with symptoms consisting of: distal bilateral lower extremity paresthesias to the mid-thighs and distal bilateral upper extremity paresthesias in the fingertips and hands, which had improved since cessation of linezolid. She developed bilateral optic neuropathy secondary to ethambutol and was found to have optic nerve swelling, left relative afferent pupillary defect, and decreased color vision after therapy with ethambutol 1200mg daily for 6 months. Optic nerve and retinal nerve fiber imaging and color vision returned to baseline after cessation of ethambutol. The patient’s current antimicrobial regimen is clofazimine, pyrazinamide, and moxifloxacin with a plan for 18 months total of anti-tuberculosis therapy.

Discussion: This case details an immunocompetent person with untreated latent TB manifesting as spinal tuberculosis with paraspinal abscesses. This case also illustrates isoniazid resistance leading to inability to use traditional RIPE therapy and demonstrates numerous examples of drug toxicities that can complicate the treatment of tuberculosis infections.

References


Disclaimer: The views expressed in this abstract are those of the author(s) and do not necessarily reflect the official policy or position of Walter Reed National Military Medical Center, Naval Medical Center Portsmouth, the Department of the Navy, Department of Defense, or the United States Government.
A Diagnosis In Sheep’s Clothing

Authors: Catey L. Harwell, MD

Introduction: Patient is a 75-year-old healthy man who presented with one day of fever and two weeks of confusion. He was recently diagnosed with systemic lupus erythematosus (SLE) in the setting of a malar rash, arthralgias, encephalopathy and a progressive 20 pound weight loss. His primary care provider started him on prednisone. On presentation, he was oriented to self only. His son provided additional history, who described a progressive “slowness” with regards to his father’s activities of daily living. In particular, he noted pain and stiffness in his hands. His symptoms were present for six months but acutely worsened over two weeks. Prior to this illness, he was operating his cattle ranch in southwestern Colorado. He also had sheep, goats, horses, and dogs. He consumed unpasteurized milk and unpurified water from nearby streams.

Case Presentation: On exam, patient was afebrile, tachycardic but hemodynamically stable. He was cachectic, and had a scaly, erythematous rash on his chest, upper back, and face in a malar distribution. He had arthritis of both wrists, metacarpophalangeal (MCP), and proximal interphalangeal (PIP) joints. His CBC with differential showed pancytopenia with a white blood cell count of 3.2x10^3/µL, hemoglobin of 10.1 g/dL and platelets 106x10^3/µL; anti-nuclear antibody (ANA) was positive at >1:2560 with dsDNA 1:40.

For the patient’s presentation, several broad categories were considered including an autoimmune or inflammatory syndrome such as SLE, malignancy, and infection. Given the history of exposure to farm animals, unpasteurized milk, and unpurified water, infectious etiologies included acute brucellosis, extrapulmonary tuberculosis, Q fever, coccidioidomycosis, Mycobacterium bovis, or Entamoeba histolytica. Computed tomography (CT) of the chest, abdomen, and pelvis were obtained – a large mass was seen in the right upper quadrant of the abdomen and a second mass in the pelvis. Magnetic resonance imaging (MRI) of the abdomen and pelvis showed complex cystic lesions with the “water-lily sign” in the liver and pelvis consistent with hydatid cyst disease secondary to Echinococcus spp.

Discussion: Initially, hydatid cyst disease was thought to be incidental and unrelated to the patient’s presentation of rash, arthralgias, and encephalopathy. On further review of the literature, Echinococcus spp. infections result in increased cytokine release, specifically IL-10, which can lead to an increased TH-2 response. Interestingly, individuals with particular HLA haplotypes are found to secrete higher levels of IL-10, and may be predisposed to developing a lupus-like presentation when they are infected with Echinococcus spp.

References

IgG Kappa Multiple Myeloma presenting as Hyperviscosity Syndrome

Authors: Christopher Nevala-Plagemann, Department of Internal Medicine, University of Utah

Introduction: Hyperviscosity syndrome (HVS) is an uncommon but potentially life threatening condition caused by increased viscosity of the blood. HVS is most commonly associated with Waldenstrom's macroglobulinemia, however, it can also be associated with polycythemia, leukemia, autoimmune disease, and, as will be discussed in this case, multiple myeloma.

Case Presentation: A 40-year-old male with no significant past medical history presented to the emergency department with a severe headache. Further questioning revealed a 2-month history of extreme fatigue, fevers, drenching night sweats, and a 2-week history of epistaxis and oral mucosal bleeding.

On presentation, patient was febrile to 38.5°C. The remainder of his vitals were within normal limits. His exam was notable for mild gingival bleeding, but was otherwise unremarkable. Workup in the emergency department revealed pancytopenia (WBC 2.6 K/mcl, Hgb 8.3 K/mcl, Plt 147 K/mcl), hyponatremia (127 mmol/L) and a significantly elevated protein gap (total protein >14.0 g/dl, albumin 2.4 g/dl). Kidney function, calcium, and the remainder of his complete metabolic panel were unremarkable. Head imaging was obtained and showed no acute intracranial pathology.

Based on these initial labs, concern for a hematologic malignancy was high. A quantitative immunoglobulin panel was obtained and revealed an IgG level of 9556. Based on his constellation of symptoms and the degree of IgG elevation, a diagnosis of hyperviscosity syndrome (HVS) was made. The patient was promptly treated with plasmapheresis and had near complete resolution of his symptoms.

Over the next few days, the remainder of his workup was completed and was notable for an elevated kappa/lambda free light chain ratio, an M spike on his SPEP, striking rouleaux formation on his blood smear, and an 80-90% monoclonal plasma cell population on bone marrow biopsy. Patient was ultimately diagnosed with IgG kappa multiple myeloma and went on to receive induction chemotherapy followed by high dose chemotherapy and stem cell rescue.

Discussion: HVS is a potentially life-threatening condition caused by increased viscosity of the blood. It is most commonly seen in patients with Waldenstrom’s Macroglobulinemia, but can be seen in multiple myeloma as was the case with this patient. In addition to the headaches, fatigue, and coagulopathy that this patient presented with, HVS can also manifest with blurred vision, diplopia, vertigo, tinnitus, somnolence and, in severe cases, seizures, stroke, priapism, and high output heart failure. Treatment involves temporizing measures with IV fluids until plasmapheresis can be initiated. It is important to note that while these patients may present with severe anemia or thrombocytopenia, transfusions should be avoided until after plasmapheresis.

While not common, internists should be able to recognize the constellation of symptoms and clinical scenarios in which hyperviscosity syndrome should be considered as prompt treatment with plasmapheresis is needed to optimally manage these patients.

References


The antibiotic that took her breath away, a case report of daptomycin induced acute eosinophilic pneumonia

Authors: Matthew Mulligan, MD; Santosh Reddy, MD; Stacy Johnson, MD

Introduction: Acute eosinophilic pneumonia (AEP) is a rare complication of daptomycin reported in case studies. As daptomycin is an increasingly used antibiotic for its activity against resistant gram-positive organisms clinicians should be able to recognize and appropriately manage AEP.

Case Presentation: A 57-year-old woman with a history of congenital hip dysplasia who had a stage one revision for an infected left total hip arthroplasty with gamella morbillorum approximately two weeks prior to presentation and discharged on a six-week course of daptomycin, presented with four days of a dry cough, progressive shortness of breath, and pleuritic chest pain. She did not have any fevers, rashes, recent travel, or history of asthma or sinus problems. On examination she was afebrile, had a room air oxygen saturation of 85%, which improved with 2-4L nasal cannula, and had bilateral crackles on pulmonary auscultation. She had a CXR with bilateral hazy opacities with peripheral and basal predominance and chest CT confirmed multifocal, multilobar parenchymal opacities. Her labs were notable for a WBC of 10.87 with 1870 absolute eosinophils, and negative ANCA’s. She was diagnosed with probable eosinophilic pneumonia secondary to daptomycin. Her daptomycin was switched to ceftriaxone, and she was started on prednisone 40mg daily. She had quick initial improvement in her dyspnea and resolution of her peripheral eosinophilia, although still required 2 L nasal cannula on discharge. Six weeks later she had significant improvement in dyspnea, was weaned from oxygen, and had near complete radiologic resolution of pulmonary opacities.

Discussion: Acute eosinophilic pneumonia (AEP) is a rare, but potentially severe, complication of daptomycin use. The mechanism of disease is unknown but is speculated to be either related to injury from drug accumulation near the alveolar surface or injury and inflammation from daptomycin binding to pulmonary surfactant. The diagnosis of acute eosinophilic pneumonia (AEP) secondary to daptomycin is typically based on the compilation of acute onset dyspnea, bronchial alveolar lavage with > 25% eosinophils or peripheral eosinophilia, new pulmonary infiltrates, fever, and concurrent exposure to daptomycin with improvement in symptoms with withdrawal of the drug. Daptomycin induced AEP must be distinguished from other causes of acute eosinophilic pneumonia including other antimicrobials, NSAIDS, helminthic and fungal infections, and autoimmune conditions such as eosinophilic granulomatosis with polyangiitis. While recovery is expected, disease can be severe occasionally requiring mechanical ventilation. In this case the patient had acute onset of dyspnea and dry cough two weeks into daptomycin therapy, peripheral eosinophilia, new diffuse pulmonary infiltrates, and rapid improvement with discontinuation of daptomycin and initiation of prednisone. Despite early improvement, she required an additional six weeks of corticosteroid therapy for further clinical and radiological resolution.

References

Pulmonary Hypertension Secondary to Carfilzomib

Authors: Julia Powelson, MD; Maryellen Antkowiak, MD; Hibba Tul Rehman, MD

Introduction: Carfilzomib is a proteasome inhibitor used for treatment of refractory multiple myeloma.

Case Presentation: The patient is a 53-year-old male with PMH refractory IgA lambda multiple myeloma on Cycle 1 Day 25 of carfilzomib who presented to the hospital with dyspnea. He reported development of chest tightness and progressive dyspnea on exertion starting shortly after his first dose of carfilzomib. He later developed orthopnea and paroxysmal nocturnal dyspnea. On presentation, physical exam was notable for elevated jugular venous pressure to 12 cmH2O and pitting edema of the bilateral lower extremities. His labs were notable for NT pro BNP 10,100. He had been evaluated pre-treatment with an echocardiogram showing normal left ventricular ejection fraction and normal right ventricular function. Echocardiogram was repeated after admission and was notable for dilated right ventricle with severely reduced systolic function, elevated pulmonary artery systolic pressure of >55 mmHg, and dilated inferior vena cava with blunted respirophasic variation. He underwent V/Q scan to rule out pulmonary embolism which was negative. He had a nuclear medicine stress test which was negative. He was treated with diuresis and was discharged. At outpatient follow up, pulmonary function testing was notable for DLCO 14.84ml/min/mmHg which was reduced at 59% of predicted. He was referred for right heart catheterization after achieving euvoolemia. Results were notable for elevated left ventricular end diastolic pressure, elevated pulmonary vascular resistance, elevated right ventricular end diastolic pressure and elevated right atrial pressure. This was felt to be consistent with mixed presentation of chronic heart failure with preserved ejection fraction with superimposed acute pulmonary hypertension. In addition to diuresis, he was started on calcium channel blockade with amlodipine. He was later started on tadalafil, which he tolerated well. His course was complicated by frequent need for intravenous fluids with chemotherapy leading to volume overload requiring intravenous diuresis. Despite this, he improved with increased exercise tolerance, resolution of volume overload and reduction in NT pro BNP levels. There was consideration of addition of endothelin receptor antagonist; however, his myeloma continued to progress and he ultimately died from acute renal failure related to progression of multiple myeloma.

Discussion: This case illustrates a rare complication of carfilzomib chemotherapy. There are several reported cases of cardiotoxicity from this medication; however, the majority of cases involve depressed left ventricular function rather than right ventricular involvement. Systemic hypertension is a common side effect of carfilzomib with a known mechanism, but isolated pulmonary hypertension as seen in this case is extremely rare. Though the association between carfilzomib and right heart failure is rare, the timing of this case with normal echocardiogram pre-treatment followed by development of severe right heart failure within three weeks of initiation of treatment strongly suggests carfilzomib as the cause of this patient’s presentation.

References

A Case of HPV related Oropharyngeal SCC in the absence of traditional risk factors: a rising clinical entity

Authors: Ahmed S. Ahmed, MD Associate Member, Sami Tahhan, MD, FACP, Eastern Virginia Medical School

Introduction: HPV-related oropharyngeal squamous cell carcinoma (OPSCC) represents a growing etiologically distinct subset of head and neck cancers with unique epidemiological, clinical, and prognostic characteristics. We report an interesting case of HPV related OPSCC in the absence of other identifiable risk factors.

Case Presentation: A 49-year-old Caucasian male with no prior history of smoking or alcohol abuse presented in 2011 with complaints of asymptomatic left-sided neck swelling. Neck CT revealed a malignant appearing complex mass in the left neck, posterior to the submandibular gland. FNA biopsies revealed proteinaceous debris and were negative for malignancy. The patient was scheduled for surgical excision but was lost to follow up.

In 2017, the patient presented again, now with complaints of a persistent left-sided neck mass. Examination revealed a firm lesion measuring approximately 3 x 4 cm at the anterior border of his left SCM.

Reimaging demonstrated a left palatine tonsillar mass with multiple multifocal ipsilateral cervical malignant appearing adenopathy concerning for metastatic malignancy from a palatine tonsil mucosal primary tumor. FNA biopsy was positive on immunohistochemistry for p16 HPV mediated OPSCC. The patient was treated with chemoradiation with a good clinical response.

Discussion: Despite the decrease in tobacco use, the incidence of oropharyngeal cancer initially remained constant and then began to rise. Public health sources describe it as an upcoming epidemic. HPV related OPSCC seems to account for this incidence trend with one study suggesting that the annual number of HPV-associated oropharyngeal cancers in the United States will overtake the incidence of invasive cervical cancer cases by 2020. As in cervical cancer, oral HPV infection appears to be a sexually-acquired disease likely through oral sex thus reflecting societal changes in sexual behavior that have occurred over time in the developed world.

HPV infection, and in particular type 16 (HPV-16), is now recognized as a significant player in the onset of OPSCC. Diagnosis is made through Immunohistochemistry for p16 which is overexpressed and highly sensitive for HPV associated OPSCC. The p16 protein functions as a tumor suppressor preventing phosphorylation of the Retinoblastoma protein (Rb) and affecting cell growth.

Compared to non-HPV OPSCC, HPV associated tumors predominantly arise in the tonsillar region or the base of the tongue, it’s more likely to present with an early-stage and usually have a better prognosis. The age of presentation tends to be younger peaking around 30 and 55 years with less exposure to the traditional risk factors of tobacco and alcohol. HPV positivity is less frequent in African Americans than in Caucasians with a threefold higher incidence in males than females. Treatment of HPV OPSCC includes a multimodality approach with chemotherapy, radiation therapy, and/or surgery with significantly better prognosis and five-year overall survival compared to HPV free OPSCC.
CNS Lymphoma Strikes a Nerve

Authors: Dachi Chelidze, MD; Nick Fuerst, MD; Nina Swiacki, DO, Faculty Author: B. Mitchell Goodman, MD

Introduction: Human Immunodeficiency Virus (HIV) can present in myriad ways, with duration of infection and degree of immunosuppression typically determining symptomatology. Likewise, patients with HIV are at risk for a number of AIDS defining illnesses (ADI) related to immunosuppression not typically seen in the general population. The most common ADI include opportunistic infections and Kaposi sarcoma, with other opportunistic malignancies representing less than two percent of ADI. We present a case of acute CN VI and VII palsy as the sentinel event prompting a diagnosis of HIV/AIDS and ultimately diffuse large B cell lymphoma (DLBCL).

Case Presentation: A 42 Y/O AAM with PMH of polysubstance abuse presented to ED with 2 weeks of worsening vertigo, headache, left facial weakness, blurred vision and new onset dysphagia. Physical exam was notable for complete left facial (VII) palsy, left abducence (VI) palsy and umbilicated facial rash. MRI of head revealed a 2.6x2.6x3.9 cm progressive brain lesion at the ponto-medullo-cerebellar junction, moderately compressing fourth ventricle. Ten days prior he had brain MRI at another hospital which showed significantly smaller 1.2x1.0 cm lesion suggestive of a demyelinating process such as multiple sclerosis. He deferred further workup at that time. Serum syphilis IgG came positive with a 1:4 titer and LP was significant for WBC 64 and lymphocytic pleocytosis. A diagnosis of neurosyphilis with gumma was entertained and the patient was placed on IV penicillin. HIV testing was positive with a CD4 count of 55 cells/mL and viral load of 100,000. EBV CSF PCR returned elevated at 119,000. The patient initially declined further invasive testing; however, he developed progressive dysphagia over the next few days and ultimately consented to brain biopsy. Brain biopsy was performed and pathology revealed EBV positive HIV associated large B-cell lymphoma. Stains for cd19, cd20, bcl2 were positive. The patient was started on IV decadron with plans for radiation and chemotherapy. To further define extent of disease, a bone marrow biopsy was performed which revealed no evidence of lymphoma. Unfortunately, he developed aspiration and respiratory distress after anesthesia for bone marrow biopsy and expired.

Discussion: The differential diagnosis for patients with cranial neuropathies is broad. Our case illustrates the importance of early recognition of immunocompromised state and tissue diagnosis to guide therapy. Diagnosis of DLBCL remains challenging because its clinical presentation could overlap with other HIV associated CNS lesions. DLBCL usually presents with supratentorial rather than brain stem lesions as with our patient. It is typically a late complication of long standing HIV infection in the setting of profound immunosuppression and rarely represents as an ADI. Brainstem involvement is characterized by aggressive nature, high rates of complications and fatality. Biopsy is always necessary to differentiate from other HIV associated lesions with markedly different treatments.
VIRGINIA CLINICAL VIGNETTE POSTER FINALIST - DANIELLE ENGSKOW, MD

Castleman disease: a rare cause of lymphadenopathy and neutropenic fever in a patient with AIDS

Authors: Danielle Grams Engskow MD, Linda Lesky MD

Introduction: Multicentric Castleman disease is a rare lymphoproliferative disorder commonly associated with HHV-8 infection and immunodeficiency. Diagnosis can be challenging because patients frequently present with non-specific symptoms such as fever, night sweats, fatigue, and lymphadenopathy.

Case presentation: A 46-year old man with a history of AIDS, COPD presented from the local jail with fever, tachycardia, three days of diarrhea, and three weeks of cough with sputum production and increasing dyspnea. He also endorsed two weeks of intermittent, subjective fevers, and night sweats. He was re-started on HAART therapy one-month prior after being off of treatment for two years. His CD4 count at that time was zero. He was admitted to jail three days prior to admission at which time he had a negative quantiferon gold. The patient was ill appearing but in no acute distress. Oral exam revealed white plaques. Lungs had diffuse inspiratory and expiratory wheezing. He had multiple, small, rubbery submandibular, cervical, and inguinal lymph nodes. Labs were notable for neutropenia, anemia, and acute kidney injury. Urinalysis was without evidence of infection. CT chest revealed RLL atelectasis, emphysema, and mediastinal, axillary, and upper abdominal lymphadenopathy. Additional testing revealed negative results for RPR, CMV antibody, HCV antibody, HBV antibodies and antigen, aspergillus antigen, toxoplasmosis IgG and IgM antibody. EVB serology was consistent with prior infection. LDH was 429. Fungitell was within normal limits. Blood cultures and fungal cultures were negative. AFB sputum smear was negative. GI PCR was positive for sapovirus. He was treated for COPD exacerbation and thrush. His diarrhea resolved with supportive care. His lymphadenopathy was attributed to IRIS. He was afebrile for the entirety of the hospitalization and was discharged on hospital day four. The patient returned one month later with fevers, chills, night sweats, and increased lymphadenopathy. He denied cough with sputum production, dyspnea, diarrhea, dysuria, or open wounds. Physical exam was notable for fever to 102.4°F and significant lymphadenopathy of the submandibular, cervical, supraclavicular, axillary, and inguinal lymph nodes. The nodes were rubbery and matted. The largest was his left inguinal lymph node, which was approximately 4cm. He was otherwise hemodynamically stable and well appearing. Biopsy of a cervical lymph node revealed multicentric Castleman disease associated with HHV-8 infection without evidence of an immunophenotypically abnormal lymphocyte population. He was initially treated with rituximab plus liposomal doxorubicin until Kaposi Sarcoma was proven negative by esophagogastroduodenoscopy and skin biopsy. He was discharged with weekly rituximab. He tolerated 5 cycles well with improvement in his lymphadenopathy, neutropenia, and anemia.

Discussion: The differential diagnosis for generalized lymphadenopathy is broad, especially when evaluating immunocompromised patients. This case demonstrates the importance of lymph node biopsy in diagnosing a rare cause of lymphadenopathy, Castleman disease.
A Rare Case of IgG4-Related Disease with Pulmonary Involvement

Authors: Ram Kommaraju DO, Jeff Carlson DO, Camelia Chirculescu MD, Haytham Adada MD, FACP

Introduction: IgG4-related disease is an increasingly recognized immune-mediated fibroinflammatory condition. The hallmarks of the disease process include an increase in serum concentration of IgG4, lymphoplasmacytic infiltration of tissues by IgG4-positive plasma cells, and tumor-like swelling of organs leading to organ failure. Involvement of nearly all body organs have been previously described, although very few cases exist in literature with pulmonary manifestations. Here, we report a case of IgG4-related lung disease (IgG4-RLD), which is a rare subset of IgG4-related disease.

Case Presentation: A 74 year old female with a history of lymphocytosis, COPD, and 50 pack-year smoking history initially presented to our hospital with acute, progressively worsening cough and shortness of breath. Review of her medical records revealed that she was previously hospitalized with an acute COPD exacerbation. A chest CT had demonstrated multiple pulmonary nodules, and several spiculated masses within the left lung fields including a 4.0 x 3.3 x 3.4 cm left upper lobe mass. The patient underwent navigational bronchoscopy with EBUS, and transbronchial biopsy of the left upper lobe lesion. Immunohistochemical staining using CD45 revealed numerous lymphocytes; CD68 revealed a small number of histiocytes, and pancytokeratin showed few trapped alveolar cells. Follow up PET scan demonstrated hypermetabolic lung masses in the left lung fields and mild lymphadenopathy. CT-guided FNA was performed, and pathology demonstrated entrapped atypical glands with fibro-inflammatory reaction. A tissue sample was sent to Pathology at the University of Michigan for review. The specimen was found to be non-cancerous, but was noted to be consistent with an inflammatory process with fibrosis. There was evidence of numerous IgG4-positive plasma cells, suggestive of possible IgG4-related lung disease. Subsequently, due to recurrent shortness of breath and COPD exacerbation, we performed a repeat chest CT which showed a new left upper lobe spiculated lung mass. After careful review of her case, including imaging, labs, and previous biopsies, the patient was diagnosed with IgG4-RLD. She was started on the appropriate treatment of high dose Prednisone therapy and had significant improvement of her symptoms.

Discussion: IgG4-RLD is a rare clinical entity leading to tumefactive and tissue-destructive lesions, which can cause respiratory failure and death. Isolated pulmonary involvement is rare among IgG4-RD, and can radiographically mimic malignancy. Four major patterns of lung involvement identified by chest CT are solid nodular, bronchovascular, alveolar interstitial, and round ground-glass opacities. However, tissue biopsy demonstrating characteristic histological features remains central to the diagnosis of IgG4-RLD. International consensus guidelines for the treatment and management of IgG4-RLD have been published, and suggest the condition responds to glucocorticoid therapy. As such, early recognition of IgG4-RLD can lead to better outcomes, and a high index of clinical suspicion is needed to confirm the diagnosis.
Arrhythmogenic Right Ventricular Cardiomyopathy: Some Things are Forever

Authors: Nicholas E. Rebhan, MD

Introduction: Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia (ARVD/C) is an inherited condition in which the myocardium is progressively replaced by fibrofatty tissue over a patient’s lifetime. It is a rare entity that most often presents in the third and fourth decade of life. This case describes the presentation of a patient with a life-threatening arrhythmic event whose initial pathology was inconsistent with ARVD/C but was ultimately found to have genetic markers consistent with the disease.

Case Presentation: 16yo F with no past medical history presented after syncope. She had reported episodes of palpitations for the previous 2 months which were often accompanied by a stressor or emotional event. On the day of presentation, she had been at cheerleading practice and experienced palpitations. Her parents arrived and took her home, where she had emesis and syncope. She awoke shortly afterward and was transported by EMS to an emergency room. EKG en route was concerning for tachycardia (HR > 250 bpm) and she was given adenosine by EMS. However, upon arrival in the ED, patient was determined to be in sustained ventricular tachycardia and was successfully cardioverted before transfer to the PICU.

Electrophysiology study showed nonsustained ventricular tachycardia and no evidence of accessory pathway. Cardiac MRI revealed patchy infiltration consistent with ARVD/C or myocarditis. Myocardial biopsy showed a “lymphocytic myocarditis in organizing phase with chronic inflammation in the interstitium composed primarily of macrophages and lymphocytes.” It was thought that ARVD/C was unlikely without evidence of fibrofatty change, however genetic testing confirmed PKP2 mutation, consistent with ARVD/C. Subcutaneous pacemaker and defibrillator was placed.

Discussion: ARVD/C was originally described in 1982 (2). It was thought to be idiopathic infiltration of the myocardium with fibrofatty tissue. It has since been found to be an inherited disease caused by desmosomal protein genetic mutations (1). The disease carries a lifetime risk of LAEs, with the highest incidence reported in the 2nd and 3rd decade of life (3). Strategies to prevent LAEs include anti-arrhythmic medication, catheter ablations, and ICD implantation. An interesting point with this case is that her biopsy did not display the expected histology of fibrofatty infiltration of the myocardium. Instead, it showed chronic inflammation, which may be consistent with the hypothesis that impaired desmosome function leads to cardiac myocyte detachment and death, which leads to inflammation and ultimately fibrofatty infiltration (4). It is also possible that her biopsy was not taken from an area of her myocardium where fibrofatty replacement had occurred. The patient described in this case presented at 16, consigning her to a lifetime of medication, procedures, LAEs, and potentially a heart transplant. This case highlights some peculiarities of ARVD/C but also illuminates the importance of lifetime follow up and excellent transition/continuity of care.

References

A case of chronic Q fever infection in a patient with a persistent endoleak after an abdominal aortic aneurysm repair

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Introduction: Chronic Coxiella burnetii (Q Fever) infection typically presents with non-specific signs and symptoms in patients with pre-existing valvular disease or rarely in those with aortic aneurysms and vascular grafts. We present a case of chronic Q fever infection in a cattle farmer with persistent back pain and weight loss who suffered a persistent endoleak after an abdominal aortic aneurysm repair.

Case Presentation: A 69 year old male is seen in clinic two months after repair of an abdominal aortic aneurysm rupture secondary to a type Ia endoleak complaining of persistent low-back pain, low grade fevers, decreased appetite and weight loss. A positron emission tomography (PET) scan is obtained and demonstrates uptake in a new left psoas collection contiguous to the persistent endoleak. He is admitted to the hospital and initial laboratory studies are significant for a normal leukocyte count, elevated sedimentation rate 49mm/hr, and an elevated c-reactive protein (CRP) 4.45mg/dL. He undergoes computed tomography (CT) guided aspiration of the psoas collection. Studies from the initial and repeat aspiration remain negative on bacteria, fungal and AFB cultures. Further workup is completed to evaluate for an infectious etiology of aortitis and reveals a positive coxiella burnetii phase II IgG antibody which reflexes to an IgG titer of 1:32. He is started on doxycycline and hydroxychloroquine for treatment of presumed chronic c burnetii prosthetic vascular graft infection. On four month follow-up patient reports complete eradication of back pain and repeat PET demonstrates resolution of the left psoas collection. He completes a total of 6 months of antibiotic therapy.

Discussion: Coxiella burnetii infection is extremely rare due to difficulty with diagnosis and availability of serologic testing. In the US, the incidence of chronic c burnetii infection is 0.3-1.0 case per million persons (1). Acute infection manifests as a self-limiting flu-like illness, pneumonia or hepatitis. Treatment is necessary to prevent chronic or persistent localized infections in those with valvular or vascular disease. Although the most common presentation of chronic infection is endocarditis (75% of cases), the incidence of Q fever endocarditis is extremely rare and is seldom reported in the U.S (2,3). Infection of aortic aneurysms or vascular grafts is the second most common presentation. Delays in diagnosis and treatment precipitate the need for multiple surgical interventions and is associated with a high mortality. (4). Our patient had improvement in his symptoms and radiographic resolution of the left psoas collection with antibiotic therapy. Given the morbidity and mortality associated with chronic Q fever infection clinicians should be aware of the clinical signs and appropriate diagnostic serology in patients with pre-existing valvular or vascular disease and exposure to coxiella burnetii.

References

**Purpuric diseases: Vasculitis or a sham?**

**Authors:** Megha Vashist, MD, Associate Member. Sami Tahhan, MD, FACP, Eastern Virginia Medical School

**Introduction:** With the many causes of purpura, determining between vascular causes versus more benign conditions is important in order to protect the patient from an expensive and unnecessary work-up. Pigmented purpuric dermatoses (PPD) is a term used to encompass a group of benign, chronic, relapsing, and remitting pigmented cutaneous lesions. The cause of PPD is unknown however it is often associated with chronic venous hypertension. Treatment remains largely uncertain.

In this case, we present a 59 year old lady who suffered from acute congestive heart failure (CHF) exacerbation, and was diagnosed to have PPD by biopsy which improved after treatment for CHF exacerbation.

**Case Presentation:** A 59 year old female with multiple comorbidities including hypertension, uncontrolled diabetes mellitus, chronic obstructive pulmonary disease, and history of gastric bypass surgery presented with complaints of dyspnea, orthopnea, and lower extremity swelling found to be secondary to acute congestive heart failure. Initial exam was significant for basilar crackles in the lungs bilaterally and lower extremity edema. Closer inspection of the lower extremities revealed irregular purpura over the dorsal aspect of bilateral feet, anterior calves, and soles of feet, which were painless and non-pruritic. The lesions started 6 months prior and had never been treated.

Workup included biopsy of the lesion in addition to serology to rule out a presumed vasculitis or infectious source. Biopsy of the lesions showed perivascular dermatitis with red cell extravasation and hemosiderin deposition, which was demonstrated with the use of iron staining. Additionally, her P-ANCA, MPO and PR-3 antibodies were negative making a P-ANCA associated vasculitis unlikely. Infectious sources were ruled out. Our patients’ characteristic lesions in addition to serology and biopsy findings were consistent with Schamberg disease- a subtype of pigmented purpuric dermatoses.

She was treated for CHF exacerbation with intravenous and oral loop diuretics and was started on a beta-blocker, ACE-Inhibitor, Statin, and Aspirin resulting in improvement of purpuric lesions suggestive of an association. Our patient was readmitted 3 months subsequent to discharge for a non-healing ulcer at the site of biopsy and was found to have right lower extremity arterial insufficiency requiring bypass.

**Discussion:** This case illustrates the importance of refraining from extensive and potentially invasive workups that could cause more harm than good to the patient. The characteristic lesions on physical exam and prompt improvement subsequent to treatment for CHF exacerbation point towards a more benign etiology such as PPD. As there is no specific treatment for Schamberg’s disease, the patient could have been spared further diagnostic work-up and resulting complications.
WASHINGTON CLINICAL VIGNETTE POSTER FINALIST - LAUREN M BENSON, MD

Itty Bitty Living Space: A Constipation Parable

Authors: Lauren Benson, MD PGY3, Internal Medicine Residency Spokane, WA

Introduction: Please see attached/emailed CT image

Case Presentation: A 62 year old female with past medical history of squamous cell bladder carcinoma presented to the emergency department for worsening abdominal pain and swelling on post operative day 7 after anterior pelvic organ exenteration with ileal conduit urinary diversion. She reported no bowel movement in 6 days. She was taking her post-op hydromorphone and docusate as prescribed. She was hemodynamically stable though visibly uncomfortable. Physical exam revealed severe scoliosis, diffuse edema and pain to palpation in her abdomen, though her urostomy and incision sites appeared intact. Her lungs were clear to auscultation with mildly diminished bases, but she required 2L nasal cannula oxygen for mild hypoxia. Cardiac exam was remarkable only for mild tachycardia. Laboratory analysis revealed BNP 61 pg/mL, AST 67, ALT 68, hemoglobin 12.3 g/dL, creatinine .49 mg/dL, and albumin 3.0 g/dL. CT scan revealed small bilateral pleural effusions with increased bibasilar atelectasis, and diffuse third spacing with ascites and anasarca. In all, this suggested possible right heart failure with sparing of the lungs relative to body edema without evidence of left heart stretch (such as elevated BNP). Upon closer review of the CT scan images and findings, a Type IV (giant) paraesophageal hernia could be seen containing the entirety of the stomach and a loop of the transverse colon which contained a large volume of stool adjacent to atelectatic lungs in the setting of thoracic restriction from severe scoliosis. This large hernia also appeared to be compressing the inferior vena cava. Upon further questioning the patient did endorse a known history of hiatus hernia which had never required surgical correction.

It is most likely that this patient’s post-operative constipation distended her hiatus hernia resulting in poor venous return to the heart as well as exacerbating her compressive atelectasis. Her mild transaminitis was likely congestive. With an intensive bowel regimen, and only two doses of 20mg of Lasix, the patient diuresed 3.5L of urine, her abdominal pain improved, and transaminitis and hypoxia resolved. She was discharged on day three.

Discussion: While bowel regimens may seem mundane, and most patients are unlikely to have such dramatic presentation or consequences to their constipation, this case provides a parable of why post-operative bowel regimens are important. Furthermore, as recent data suggest that docusate is no more effective than placebo, it is important to re-focus our prescribing practices (and automated order sets) toward more efficacious medications such as polyethylene glycol. Finally, as internists, we too often leave contemplating the three-dimensional anatomic relationship of organs to the surgeons, yet in this case the malfunction was more structural than dysfunctional.

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Important Questions to Diagnose Intractable Chronic Abdominal Pain

Authors: Paula Morzenti, MD PGY2 (Associate), Patricia Short, MD (Fellow)

Introduction: Chronic abdominal pain can be difficult to distinguish between abdominal wall (somatic pain) and intra-abdominal cavity (visceral pain) in the clinical setting. We present a case of a 19 year old active duty female with chronic abdominal pain for 6 months ultimately diagnosed with abdominal cutaneous nerve entrapment syndrome. Careful history and physical examination were paramount to establishing the diagnosis.

Case Presentation: A 19 year old female without significant past medical history referred to clinic endorsing a 6 month history of constant left upper quadrant abdominal pain associated with daily nausea and emesis. Review of systems otherwise unremarkable. Pain began 6 month prior when she was admitted for pyelonephritis with classic symptoms of LUQ pain, dysuria, frequency, nausea and vomiting. While the urinary symptoms resolved, she noted the LUQ pain persisted. A follow-up evaluation revealed a positive chlamydia culture. Despite appropriate antibiotic treatment, the pain persisted. Abdominal ultrasound and CT abdomen/pelvis were unrevealing and patient was given a different antibiotic for presumed failure of pyelonephritis therapy. Patient was seen 2 months later for ongoing symptoms and GU exam revealed right adnexal tenderness concerning for pelvic inflammatory disease, of note transvaginal ultrasound was unremarkable. Patient managed as an outpatient for PID but returned to the ED one day later with continued abdominal pain and was admitted for suspected outpatient treatment failure. General surgery was consulted with recommendations for exploratory laparotomy which patient declined. Ultimately, patient was treated with antibiotics again for presumed pyelonephritis and placed on narcotic pain medications. Despite multiple courses of antibiotics, pain, nausea and emesis remained essentially unchanged. After discharge, patient was referred to the Internal Medicine Consult service for further evaluation. Physical exam notable for focal LUQ tenderness that worsened with straight leg raise, consistent with a positive Carnett’s sign. Patient was diagnosed with anterior cutaneous nerve entrapment and scheduled for trigger point injection.

Discussion: Recent studies demonstrate the abdominal wall is the cause of chronic abdominal pain in up to 30% of patients with chronic abdominal pain and nearly 2% of patients in the ED endorsing acute abdominal pain. Misdiagnosis and delayed diagnosis lead to escalated health care costs and potentially unnecessary procedures and harm to patients. Carnett’s test has a sensitivity and specificity that approach 78% and 88% respectively alone and up to 85% and 97% respectively when used in combination with additional diagnostic criteria. This test is easy and inexpensive and therefore an important consideration in the assessment of chronic abdominal pain. Anterior cutaneous nerve entrapment has a much greater incidence than initially thought and therefore is important to consider in patients presenting with both acute and chronic abdominal pain.

References

Sliding back: retrospective histopathologic diagnosis of cervicothoracic pain

Authors: Stephen Slade, MD, Department of Graduate Medical Education, Virginia Mason Medical Center, Seattle, Washington, Erin Bauer, MD, Section of Rheumatology, Virginia Mason Medical Center, Seattle, Washington, Amish Dave, MD, MPH, Section of Rheumatology, Virginia Mason Medical Center, Seattle, Washington

Introduction: While back pain is among the most common chief complaints and often musculoskeletal in origin, more sinister and exotic etiologies must be identified, as in this case.

Case Presentation: A 51-year-old male presented with 2 months of isolated upper back pain initially suspected secondary to overuse and muscular strain. Over the next 3 months, his pain progressively worsened, and he developed lower extremity dysesthesias and subjective weakness despite a normal neurological exam. Lab workup was largely non-revealing with notably normal muscle enzymes, C-reactive protein, and urinalysis with negative anti-nuclear antibody (ANA) multiplex and HLA-B27. Vitamin B12 was low at 245pg/mL, and erythrocyte sedimentation rate was slightly elevated at 30mm/h. He had positive perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA) with myeloperoxidase (MPO) level 3.3U (1.1U 14 months later with persistent p-ANCA) without evidence of end-organ damage to suggest vasculitis.

Three months following initial presentation, magnetic resonance imaging (MRI) revealed a C7-T5 epidural mass and normal brain, prompting neurosurgical evaluation and recommendation for laminectomy with evacuation of suspected hematoma. He initially deferred surgery because his symptoms fully and promptly resolved following a 5-day course of prednisone 40mg. His symptoms recurred within 2 months with corresponding neurologic changes, and 5 months later he underwent T4-T5 laminectomy with biopsy of a mass diffusely adherent to dura mater. Pathology showed fibrotic tissue of unclear etiology with polyclonal lymphoid infiltrate, which is inconsistent with B cell lymphoproliferative disorder, and showed no malignant cells, vasculitis, or granulomas.

In the 2 years following initial back pain onset he received prescriptions across 4 departments for 11 corticosteroid courses, each lasting 5-7 days. He then presented to rheumatology clinic for worsening back pain and headaches relieved only with corticosteroids. Repeat spinal MRI demonstrated epidural thickening and severe canal stenosis with suspected cord compression. This progression prompted IgG4 immunostaining on biopsies from the prior year, which revealed focally dense IgG4-positive plasma cells, up to 29/hpf, consistent with spinal IgG4-related hypertrophic pachymeningitis. Since starting a steroid-sparing disease-modifying anti-rheumatic drug with prednisone taper, changes in his symptoms and neurologic exam have shown promising results.

Discussion: IgG4-related disease is an inflammatory disorder characterized by particular histopathology: 1) dense lymphoplasmacytic infiltrate, 2) fibrosis with at least focal storiform pattern, and 3) obliteratorive phlebitis. Diagnosis in this case required multidisciplinary evaluation with particular value from retrospective histopathologic analysis. Recognition of this disease entity may be obscured by its multi-system protean manifestations, including sialoadenitis, interstitial pneumonitis, pancreatitis, tubulointerstitial nephritis, and retroperitoneal fibrosis. Spinal IgG4-related hypertrophic pachymeningitis is a rare presentation of IgG4-related disease. To our knowledge, there have previously been only 10 total reported cases and only 1 other with concomitant p-ANCA/MPO positivity. If diagnosed early enough, treatment may prevent progression to permanent neurological impairment and functional disability.
An Uncommon Manifestation of IgG4 Related Disease: Laryngeal Involvement

Authors: Azfar Syed, DO (ACP Associate); Peter Henning, DO, Madigan Army Medical Center

Introduction: IgG4 related disease (IgG4 RD) is an immune mediated fibro inflammatory condition caused by an accumulation of IgG4 antibodies produced by B-cell plasmacytes causing dysfunction in target organs. It is a relatively recently recognized condition which encompasses several previously defined organ-specific conditions which share this common pathophysiology. Among the more common presentations are autoimmune pancreatitis, lymphadenopathy, and salivary gland involvement. However, the manifestations of IgG4 RD are protean and it can affect multiple organ systems, frequently simultaneously. The diagnosis is suggested by clinical symptoms and labs which typically demonstrate elevated serum IgG4 levels. Tissue biopsy with characteristic increased IgG4 plasma cells confirms the diagnosis. Here, we report a unique case of IgG4 disease affecting the larynx.

Case Presentation: A 69-year-old male with a history IgG4 autoimmune pancreatitis diagnosed two years prior presented with progressive upper airway symptoms. He initially endorsed cough. With time, he developed hoarseness and severe episodes of coughing with copious secretions and inability to breath. Direct laryngoscopy showed inflammation of the epiglottis and surrounding tissues and overt vocal cord dysfunction. His symptoms were ascribed to reflux disease. He was started on a regimen of high dose proton pump inhibitor, H-2 blocker and sucralfate. His vocal cord dysfunction continued to worsen, and he underwent an upper endoscopy which was unremarkable. He also had a normal barium swallow study which ruled out acid reflux. A PET scan was performed which showed PET avidity in lymph nodes as well as his aorta. Due to concern for progressive IgG4 related aortitis, he was treated with a combination of moderate dose systemic corticosteroids and rituximab infusions with resolutions of his symptoms. Repeat direct laryngoscopy was performed which revealed no further evidence of laryngeal inflammation. Given his significant clinical improvement in response to this therapy and his PET scan findings we believe this to be a case of IgG4 laryngeal disease.

Discussion: There have only been a handful reported cases in the literature of IgG4 disease affecting the larynx. This patient had previous biopsy confirmed IgG4 related disease of his pancreas. The rapid improvement of the patient’s symptoms after being treated for IgG4 disease and the resolution of laryngeal inflammation strongly suggest this case being a laryngeal manifestation of this disease process. IgG4 RD of the larynx is an underreported manifestation that should be considered in patients with upper airway symptoms, especially if symptoms are refractory or if there is a history of IgG4 or additional symptoms suggestive of IgG4.

The views expressed are those of the author(s) and do not reflect the official policy of the Department of the Army, the Department of Defense or the U.S. Government.

References

Treatment of Cardiac Arrhythmias with Electrical Cardioversion in Pregnancy

Authors: Catherine Adams MD, Hisham Hirzallah MD, Julia Parsons DO, Ellen Thompson MD, Department of Internal Medicine, Department of Cardiology, Marshall University, WV

Introduction: Physiological changes in pregnancy can result in tachyarrhythmias in previously asymptomatic Wolf Parkinson White (WPW) patients. Sustained symptomatic arrhythmias can result in hemodynamic compromise in mother and fetus; therefore, rapid intervention is crucial. Electrical cardioversion (ECV) is utilized in life-threatening situations when the patient is hemodynamically unstable. Literature review indicates there are no large-scale studies concerning the safety and efficacy of ECV in pregnant populations. This case report describes the effective use of ECV during pregnancy.

Case Presentation: A 26-year-old white female G1P0 at 33 weeks of gestation with a past medical history significant for asymptomatic WPW syndrome presented with: sudden onset of shortness of breath, sustained palpitations, dizziness and chest tightness. Her heart rate was 180-220 bpm and blood pressure was 84/60 mmHg with MAP of 68 mmHg. On physical examination, gravid uterus above the umbilicus was noted. Initial serial EKGs showed wide complex irregular tachyarrhythmia and atrial fibrillation with RVR. Her laboratory work up was normal. She was evaluated by a team consisting of Cardiology, OB/GYN and Electrophysiology. She underwent cardioversion with synchronized 150J DC shock under sedation without maternal or fetal complications. Post-procedure EKG showed sinus rhythm with heart rate of 90 bpm, short PR interval and delta wave. She was admitted for observation overnight and she was started on flecainide (50 mg daily) to prevent further tachyarrhythmias. On the following day, she was discharged without additional sequelae with plans for post-partum ablation.

Discussion: WPW syndrome was first described by Wolf, Parkinson and White in 1930. It is a preexcitation syndrome due to an anomalous pathway bypassing the AV node. The overall incidence of arrhythmia resulting in WPW syndrome is reported to be up to 1% per year. However, the exact incidence of tachyarrhythmias in pregnant WPW syndrome patients is unknown. In 1961, Gleicher et al suggested that pregnancy may facilitate onset of tachyarrhythmias in patients with asymptomatic pre-excitation syndrome. Tachyarrhythmias in pregnancy can result in hemodynamic compromise to the fetus causing hydrops fetalis. It is also reported to cause premature delivery and intrauterine growth restriction. Therefore, rapid conversion to sinus rhythm is crucial for the wellbeing of both mother and fetus. Trials of medical cardioversion were reportedly unsuccessful in the past and given the special circumstances in our case where the patient was hemodynamically unstable, rapid management with electrical cardioversion under sedation was considered.

Conclusion: Pregnancy predisposes tachyarrhythmias in previously asymptomatic WPW syndrome patients. Electrical cardioversion can be considered a safe and effective treatment.
WEST VIRGINIA CLINICAL VIGNETTE POSTER FINALIST - ANDREA STARK, DO

Fractured Inferior Vena Cava Filter Strut Migration to Pulmonary Arteries

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Brandon Rose, DO, Assistant Professor, Department of Medicine, Charleston Area Medical Center/West Virginia University-Charleston Division, Charleston, WV.

Introduction: Current guidelines recommend an inferior vena cava (IVC) filter in patients with an acute deep vein thrombosis (DVT) in the setting of an anticoagulation contraindication. However, IVC filters are associated with rare life-threatening complications such as perforation, fracture, and migration, although occurrence is rare. IVC filter strut migration to the coronary and pulmonary arteries has been described, but the absolute incidence is unknown.

Case Presentation: A 19 year-old Caucasian female with a past medical history of tobacco use and left iliofemoral DVT secondary to May-Thurner Syndrome presented to the emergency department (ED) with chief complaints of chest pain and shortness of breath. In the ED, she described substernal sharp, constant, and pleuritic chest pain that was unrelieved by aspirin or nitroglycerin. Her vitals were stable aside from mild tachycardia. Chest x-ray evidenced curvilinear foreign bodies in bilateral lungs bases. CTA chest was negative for pulmonary embolism (PE), but confirmed linear foreign bodies in bilateral lower lobe pulmonary artery branches. Radiologist confirmed foreign bodies were strut fragments from her IVC filter. Six years prior, due to Coumadin noncompliance with resultant subtherapeutic INR values, a Celect IVC filter had been placed. Temporary IVC use was intended but retrieval had been unsuccessful approximately one month after placement.

Discussion: This case represents a rare, life-threatening IVC filter complication. The Celect IVC filter has since been removed from the market with numerous lawsuits pending. In 2014, the FDA conducted a quantitative decision analysis regarding the safety of IVC filter use and released a statement recommending patients with retrievable IVC filters, in whom the transient risk of PE has passed, should have their IVC filters removed. Likewise, the PREPIC studies failed to show either a reduction in mortality or recurrent symptomatic PE. Despite all of this, use of retrievable IVC filters has been increasing. We caution that unless future studies demonstrate long-term efficacy without life-threatening complications, IVC filters should be reserved only as a last resort option.
Pericardial Metastasis from an Aggressive Anal Squamous Cell Carcinoma

Authors: Muhammad Yasin, MD1, Sean Parker1, Mark Radow, MD1, Moussa Sissoko, MD1, 1. Charleston Area Medical Center, Charleston, WV

Introduction: Metastatic disease of the pericardium marks an underreported and potentially serious complication of various malignancies. Although breast and lung primary tumors represent the large majority of pericardial metastases, distant lymphatic spread of gastrointestinal tumors have been reported in the literature. We present a unique and first reported case of a fatal malignant pericardial effusion due to metastatic spread of an anal squamous carcinoma to the pericardium.

Case Presentation: A 63-year-old female with stage III squamous cell anal carcinoma (T4N2) received treatment with a regiment of 5-fluorouracil, mitomycin C, and adjuvant pelvic radiation therapy. Six months following treatment completion the patient developed hemodynamic compromise, pulsus paradoxus, and both radiographic and echocardiographic evidence of a large pericardial effusion consistent with cardiac tamponade. The effusion resolved following an emergent pericardiocentesis and consequent video assisted surgical formation of a pericardial window. Pathologic evaluation of the pericardium revealed highly aggressive p16 positive dysplastic cells consistent with a squamous cell anal primary. Two months later the patient passed away after multiple complications associated with metastatic involvement of the pericardium, liver, and aortocaval lymph nodes.

Discussion: Regional lymph nodes, liver, and lung are the most common sites of metastasis from anal cancer. However, infrequent sites of metastases have also been reported. From our literature review, this is the first case ever reported with pericardial metastasis with anal primary. One reason could be that cardiac or pericardial metastasis are usually underreported as they usually remain undiagnosed unless very advanced. Autopsy reports have suggested around 10-18% cardiac metastasis with colon to be primary in 1-7%. Proposed mechanism of pericardial involvement include network of blood and lymphatic flow in the region. The diagnosis of malignant pericardial effusion is made by means of pericardiocentesis, pericardioscopy, or both. Results of cytologic studies are positive in 80%–90% of patients with malignant pericardial effusions. A malignant pericardial effusion is associated with decreased survival, but therapeutic options include creation of a pericardial window (subxiphoid pericardiostomy), pericardial sclerosis through a small-bore catheter and radiation therapy.

References

AN ATYPICAL CASE OF ACUTE RENAL FAILURE

Authors: Lauren Banaszak, MD; Christine Seibert, MD

Introduction: Hemolytic uremic syndrome (HUS) is characterized by hemolytic anemia, thrombocytopenia, and renal failure. Atypical or complement-mediated HUS is caused by dysregulation of the alternative complement pathway. We present a case of hereditary atypical HUS caused by a gain-of-function mutation in the gene encoding complement factor 3 (C3).

Case Presentation: A 48-year old man presented with a 4-day history of abdominal pain, nausea, vomiting, and diarrhea. His medical history was significant for a Zenker’s diverticulum; he had no prior surgeries, no known allergies, and took no medications. Physical exam was remarkable for hypertension but was otherwise normal. Initial laboratory evaluation was significant for mild hyponatremia, mild hypochloremia, an anion gap metabolic acidosis, BUN 96 mg/dL, creatinine 14.2 mg/dL, normal WBC with normal differential, hemoglobin 10.3 g/dL, and platelets 63 K/ul. Urinalysis showed large blood but only 3-5 RBCs and a bland sediment. Subsequent workup revealed elevated total bilirubin, elevated LDH, undetectable haptoglobin, elevated reticulocyte count, negative direct Coomb’s, and peripheral smear with many schistocytes. His stool was negative for Shiga toxin and ADAMTS13 activity was within the normal range. The patient was urgently started on intermittent dialysis and plasma exchange for presumed atypical HUS. A kidney biopsy was performed which revealed evidence of thrombotic microangiopathy. His kidney function and hemolysis failed to improve, so the patient was initiated on eculizumab. With this, plasma exchange was ultimately discontinued but he remained dialysis-dependent. Genetic studies later revealed elevated levels of soluble C5b-C9 membrane attack complex (MAC) and a heterozygous C3 p.Lys65Gln mutation associated with hereditary atypical HUS.

Discussion: Atypical HUS is characterized by microangiopathic hemolytic anemia, thrombocytopenia, and renal failure. The absence of Shiga toxin-mediated illness distinguishes this from typical HUS, and the lack of ADAMTS13 deficiency excludes thrombotic thrombocytopenic purpura (TTP). Atypical HUS is caused by increased activation of the alternative complement pathway, leading to excessive MAC formation with subsequent endothelial cell damage and thrombotic microangiopathy. There are regulatory proteins that prevent over-activation of the alternative complement pathway. Hereditary atypical HUS is caused by a loss-of-function genetic mutation in a complement regulatory protein or more rarely an activating mutation involving a complement effector gene. The C3 mutation detected in this patient was predicted to lead to decreased binding of the regulatory protein complement factor H, resulting in impaired inactivation of C3b and increased formation of MAC. Eculizumab is an anti-complement agent that targets C5 and prevents assembly of MAC; this can be used to supplement plasma exchange in the treatment of atypical HUS.

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**Wisconsin Clinical Vignette Poster Finalist - Benjamin Lipanot, MD**

Idiopathic Fibrosing Mediastinitis: An Insidious Presentation of a Rare Disease

*Authors*: Benjamin Lipanot, MD, Gabriel Ryan, MD, Kelly Staricha

**Introduction**: Fibrosing mediastinitis is a rare disease most often associated with prior or concurrent *Histoplasma* infection. Idiopathic fibrosing mediastinitis is an even more rare subset of pathology with no clear etiology. There are only dozens of reported cases in the United States.

**Case Presentation**: A 19-year-old man presented to the emergency department with recurrent left axillary abscesses. He suddenly developed anxiety with palpitations and electrocardiogram demonstrated supraventricular tachycardia. His medical history included Crohn's disease with regular infliximab infusions and recurrent draining abscesses over his limbs, buttocks, and trunk.

He required a diltiazem drip to control heart rates. Chest X-ray revealed right upper lobe infiltrate. He was started on doxycycline and ceftriaxone to cover community acquired pneumonia and epidermal abscesses, and was admitted to the medical intensive care unit for further management of tachycardia and presumed pneumonia. Physical exam revealed regular rhythm tachycardia, normal lung findings, and multiple small scars on legs, buttocks, and trunk with draining abscess in left axilla and right thigh. He transitioned to oral diltiazem the following day with adequate rate control. Echocardiogram unexpectedly revealed an avascular intra-atrial mass with irregular borders extending between the atria and moderate pericardial effusion. CT chest, abdomen, and pelvis with contrast showed an 8.1x5.7x14.8 cm homogeneous soft tissue mass in the medial mediastinum. The superior vena cava and right pulmonary artery and veins were circumferentially encased and significantly narrowed. Mass effect was noted on right and left atria, aortic arch, and inferior vena cava. The trachea, mainstem bronchi, and distal right lung bronchi were encased without mass effect. Additionally, multiple irregular nodules in the right lung were seen. Quantiferon gold was negative. Cardiac MRI confirmed CT findings as described above, and ventricular function was not affected. He was transferred to the acute medical floor.

Superficial abscesses cultures grew *Actinomyces turicensis* and *Actinomyces neuii*, which were an unlikely cause of mediastinal mass based on imaging and subsequent pathology. He completed oral antibiotic therapy, and Infliximab was discontinued due to recurrent abscesses. EBUS with biopsy of mass was nondiagnostic but negative for malignancy. Flow cytometry of serum was negative for malignancy. *Histoplasma* urine antigen, and *Histoplasma, Blastomyces*, and *Aspergillus* serum antibodies were negative. Thoracic surgery was consulted for mediastinoscopy. Tissue and sputum cultures were negative for bacteria, fungi and mycobacteria. Open biopsy results showed dense connective tissue without signs of malignancy, consistent with the diagnosis of fibrosing mediastinitis.

**Discussion**: Fibrosis mediastinitis has no established effective medical treatment. Surgical interventions for compression of vascular or airway structures have only been shown to be temporizing.

This case further illustrated the often insidious onset of this disease with presentation typically due to significant compression of mediastinal structures. Specifically, supraventricular tachycardia prompted evaluation with an echocardiogram which led to this diagnosis.
A Challenging Case of Chronic Tophaceous Gout

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Introduction: At least 8.3 million US adults are affected by gout. The typical characteristics of gout include hyperuricemia and precipitation of monosodium urate crystals, which may cause acute inflammatory arthritis, articular erosions, tophi, and potentially uric acid renal stones and nephropathy. Of the affected population, approximately 25,000-100,000 individuals are projected to be refractory to first-line management with oral urate lowering therapy (ULT). Pegloticase is a porcine recombinant, polyethylene glycol-conjugated uricase approved for treatment of refractory gout in patients who have failed conventional ULT.

Case Presentation: A 28-year-old male was referred to rheumatology clinic for chronic tophaceous gout with cutaneous nodules. Prior to presentation he was initiated on urate lowering therapy with allopurinol and colchicine-probenecid, but he had not noticed any significant changes. Physical examination revealed tophaceous gout affecting bilateral hands, elbows, and feet. Tophi on left 1st toe and right 5th toe were noted to be oozing. One month prior to evaluation he underwent radiographic imaging of hands and feet, which showed progression of severe gout with extensive destructive erosive changes. The patient’s serum uric acid concentration (sUA) had decreased from 10.1 mg/dl to 7.7 mg/dl after beginning ULT and diet modification. He also proceeded with excision of right thumb gouty tophus with pinning of the IP joint.

Due to the extent and magnitude of tophi with erosive/destructive changes in various joints, pegloticase (Krystexxa) infusions were recommended. He received his first pegloticase infusion after sUA resulted at 10.5 mg/dl, and it was determined that if subsequent value is higher than baseline, infusions would stop. Patient tolerated the first treatment well. However, two weeks later, he developed symptoms of throat and chest tightness, nasal congestion and conjunctival injection approximately thirty-five minutes after his second infusion was started. Medication was promptly stopped and patient received fluids, diphenhydramine and hydrocortisone for the suspected infusion reaction (IR).

Discussion: Refractory gout refers to persistent arthritic symptoms, tophi, or the inability to achieve sUA below the therapeutic goal of 6.0 mg/dl despite the use of conventional ULT. The American College of Rheumatology (ACR) and European League against Rheumatism (EULAR) recommendations advise the use of pegloticase in patients with severe gout and impaired quality of life, or in whom the disease cannot be adequately treated with other available drugs at the maximal dosages. Pegloticase has shown benefit in clinical trials and open-label extensions with demonstration of reduction in plasma urate levels and tophi resolution; improvements in quality of life and physical function were also seen. Nonetheless, the medication also has potential to provoke immunogenicity, triggering some patients to develop drug antibodies with loss of efficacy and development of IRs. Additionally, it is recommended to discontinue pegloticase in patients with two sequential sUA greater than 6.0 mg/dl to reduce risk of IRs.
Suppurative Thrombophlebitis of the Facial Vein: A Variant of Lemierre's Syndrome

Authors: Xiaoxiao Qian, MD; Fisseha Ibsa, MD

Introduction: Lemierre’s syndrome refers to infectious thrombophlebitis that is commonly caused by Fusobacterium Necrophorum. The condition always affects the jugular vein and is frequently preceded by pharyngitis, usually in association with tonsil or peritonsillar involvement. Patients typically present acutely with fever (>39°C) and rigors, often accompanied by respiratory distress. Septic emboli to the lung are common.

Case Presentation: This case involved a 33-year-old Hispanic female who presented with a six-day history of fever, chills, nausea, vomiting and profound diarrhea. Patient was hypotensive, tachycardic, tachypneic and afebrile. She was in mild distress with dry oral mucus and enlarged submandibular lymph nodes. The remaining of the physical examination was unremarkable. Patient's blood pressure did not improve with aggressive fluid resuscitation. She was placed on pressors for blood pressure maintenance and cefepime and vancomycin for presumed septic shock. Initial work up was significant for mild leukocytosis with left shift, acute kidney injury and hypokalemia. Her septic shock and gastrointestinal symptoms resolved with above treatment on the second day. Antibiotics were discontinued as no source of bacterial infection were identified with initial work up and her symptoms were considered to be caused by acute viral infection. However, she continued to have high grade fever and developed worsening leukocytosis. Meanwhile, she complained of increased swelling of her right neck and progressive shortness of breath. She reported a history of dental abscess on the right side three months ago but denied any current symptoms. Her initial blood cultures obtained in emergency department grew Fusobacterium necrophorum on hospital day four. Although she did not have the typical history of pharyngitis, given the submandibular lymph nodes enlargement and blood cultures growing Fusobacterium species, Lemierre’s syndrome was suspected and Computed tomography (CT) of neck, chest, abdomen, pelvis was obtained to investigate the source of infection and possible systemic septic emboli. The imaging indicated purulent thrombophlebitis of the right facial vein and periapical abscess of the right maxillary incisor, where a branch of the right facial vein was draining from. In addition, there were cavitary lesions in her right upper lobe of lung, which were suspicious for septic emboli. Patient was considered to have a variant of Lemierre's syndrome. She was on intravenous metronidazole until her fever subsided and respiratory distress resolved. She continued oral amoxicillin/clavulanic acid for three weeks.

Discussion: This case illustrates a variant of Lemierre’s syndrome that does not have preceding pharyngitis, involves the facial vein instead of the jugular vein, and initially presents with prominent gastrointestinal symptoms. Recognizing atypical presentations of Lemierre’s syndrome is essential as prompt treatment is the key to prevent serious systemic complications.

References

To Cut or Not to Cut? A Case of Isolated Pulmonic Valve Endocarditis

Authors: Aravind Seetharaman, DO Internal Medicine, Katie E. Cohen, MD Internal Medicine, Sarah Nickoloff, MD. Assistant Professor, Internal Medicine.

Introduction: Isolated pulmonic valve (PV) infective endocarditis (IE) is a very rare finding. There are clearly defined criteria for surgery in left-sided IE. However, guidelines for surgery in right-sided IE are much less clear.

Case Presentation: A 70-year-old male with a history of alcohol abuse and hypertension presented with severe dyspnea, 30-pound weight loss, fever, and a new heart murmur. Blood cultures were positive for Enterococcus faecalis. Trans-thoracic echocardiogram (TTE) revealed a mobile vegetation on his PV. Confirmatory trans-esophageal echocardiogram (TEE) revealed a 4.0 x 1.0 cm mobile mass attached to the base of the right cusp of the PV, associated with moderate pulmonic regurgitation. Clinical findings were consistent with isolated pulmonic valve infective endocarditis. Treatment with ampicillin and ceftriaxone was initiated. Cardiothoracic surgery (CTS) was consulted but declined intervention due to his multiple medical comorbidities. They felt that he had stable cardiac function, as there was no evidence of heart block on ECG, valvular abscess formation or heart failure. They recommended cardiac MRI for further characterization, which showed destruction of the left cusp, along with cusp prolapse into and out of the RV outflow tract. Despite ongoing treatment with IV antibiotics, the patient had recurrent febrile episodes associated with hemodynamic instability. Chest CT revealed new soft-tissue densities with surrounding ground-glass infiltrates in the right lung fields. These findings were most concerning for new septic emboli. He was transferred to an affiliated hospital for a second opinion by CTS, who also felt he should continue with medical management. After completing six weeks of IV antibiotics, surveillance blood cultures appeared sterile. However, repeat TTE showed a new mobile mass (2.7 x 0.6 cm) attached to the PV, along with a smaller previously imaged vegetation (1.8 x 0.3 cm), associated with severe pulmonic regurgitation and moderately enlarged RV. Repeat blood cultures remained sterile and patient was closely monitored off antibiotics. Surveillance TTE two weeks later showed further enlargement of prior echodensities. CTS once again evaluated the patient, but continued to recommend medical management and serial TTEs.

Discussion: Initial treatment of PV endocarditis is usually medical. Our patient’s high-risk features such as vegetation size >20 mm, recurrent pulmonary emboli, severe pulmonic regurgitation and development of new, additional vegetations while on broad-spectrum antibiotics made a compelling case for surgical intervention. However, CTS felt that risk of surgery outweighed benefits, and therefore, our patient was managed medically. Right-sided heart valve infections are thought to be more responsive to antibiotics and better tolerated than mitral or aortic valve infections. However, further research is needed to establish definitive guidelines for surgical management of PV endocarditis.

References

Adult T-cell Leukemia-Lymphoma Presenting as Severe Hypercalcemia

Authors: Joy Tang, MD, Medical College of Wisconsin, Milwaukee, WI

Introduction: Adult T-cell leukemia-lymphoma (ATL) is an extremely rare peripheral T-cell neoplasm that is associated with human T-lymphotropic virus type 1 (HTLV-1). ATL generally has a poor prognosis with shorter overall survival compared to other peripheral T-cell lymphomas. We present a rare case of HTLV-1 associated ATL that presented with severe hypercalcemia.

Case Presentation: Patient is a 66-year-old female with history of hypertension who presented with altered mental status. On presentation, vitals were significant for tachycardia. Labs were significant for calcium of 17.4, white blood cell count of 155 10e3/µ with 94% lymphocytes, hemoglobin 8.3 g/dL and platelets 111 10e3/µ. Parathyroid hormone level was low. She was treated with IV fluids and pamidronate with improvement of her hypercalcemia and mental status. Hospital course was further complicated by acute hypoxemic respiratory failure requiring two days of mechanical ventilation. Chest imaging was concerning for acute pulmonary edema and pneumonia. She was diuresed with IV Lasix and treated with IV Zosyn and Vancomycin with improvement in respiratory status. Patient underwent a bone marrow biopsy that showed t-cell lymphoproliferative disorder. CT chest/abdomen/pelvis showed multiple enlarged lymph nodes in the pelvis, upper abdomen and retroperitoneum. Nuclear medicine bone scan and CT head revealed lytic lesions in the calvarium. Bone marrow slides sent for pathology revealed mature T-cell lymphoproliferative disorder occupying 30% of the marrow space with leukemic phase. She was then found to be positive for HTLV-1, making her diagnosis consistent with ATL. She was given dexamethasone and started on cyclophosphamide, vincristine and prednisone for chemotherapy.

Discussion: ATL is an aggressive post-thymic lymphoproliferative neoplasm of T-cells associated with HTLV-1 that generally has a poor prognosis. Even with chemotherapy, the median survival time is approximately 13 months. Infection with HTLV-1 is typically endemic to Japan, the Caribbean, western Africa and southeastern United States. Development of ATL occurs in <5% of patients with HTLV-11. In the U.S., incidence of ATL is approximately 0.05 cases per 100,000 people3. Diagnosis is made from identification of at least 5% abnormal T-lymphocytes in the peripheral blood and confirmation of HTLV-1 infection. Clinical features can vary, but include circulating leukemia cells, lymphadenopathy, hepatosplenomegaly, lytic bone lesions and opportunistic infection. Severe hypercalcemia (serum calcium >14 mg/dL) can be seen in >80% of ATL. Many factors have been implicated in the mechanism for hypercalcemia, including interleukin-1, transforming growth factor-β and parathyroid-related protein (PTHrP). Among those factors, PTHrP is considered to play a significant role by stimulating osteoclasts and thus increasing bone resorption as ATL cells typically express large amounts of PTHrP4. It is important to aggressively treat severe hypercalcemia as it has been associated with early mortality2. Physicians should also be aware of the clinical features of ATL to improve early diagnosis and involve hematology/oncology early for treatment.

References
